Dermatology 's hidden treasures

Dr. Hany Salah Soliman

First edition 2025

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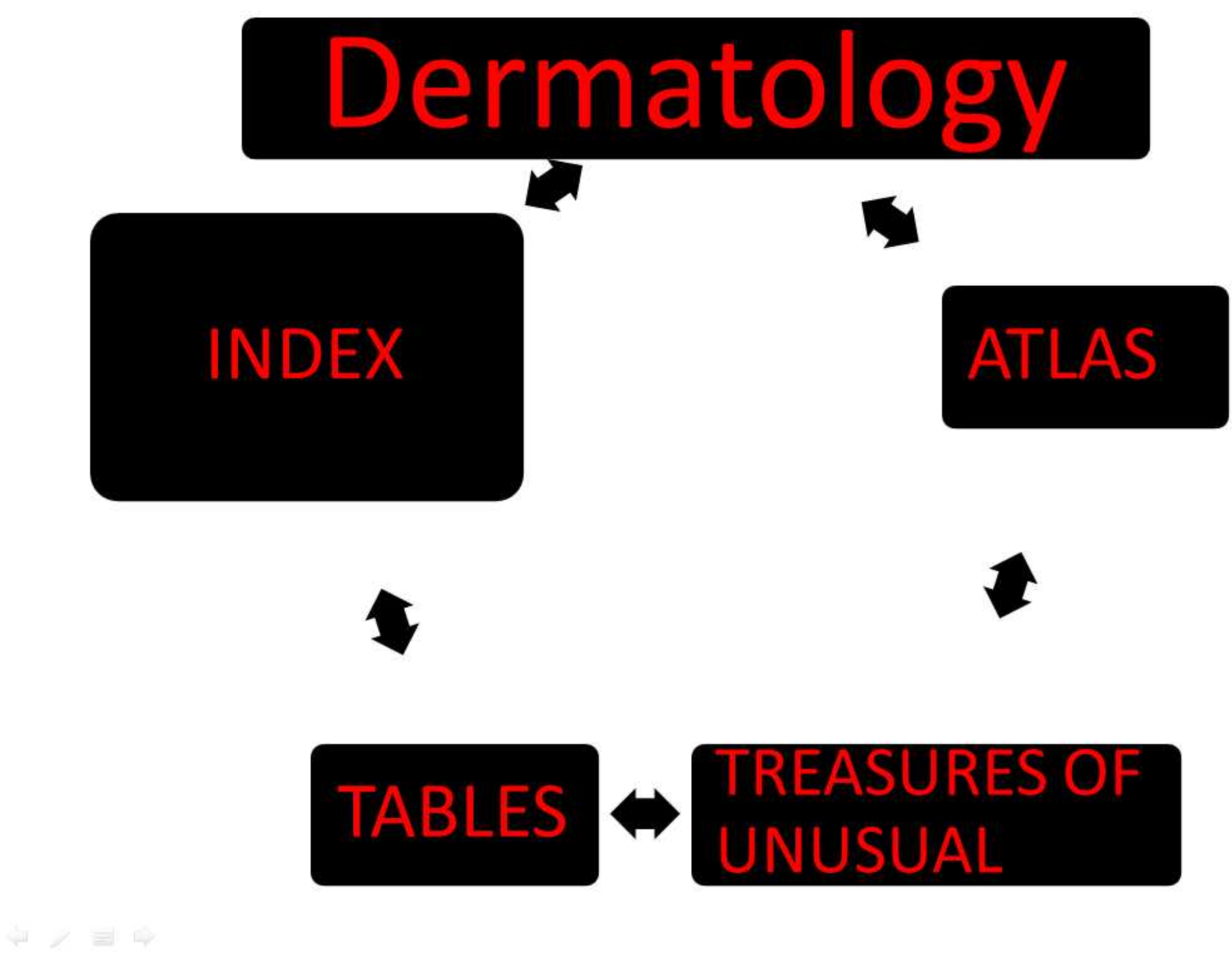
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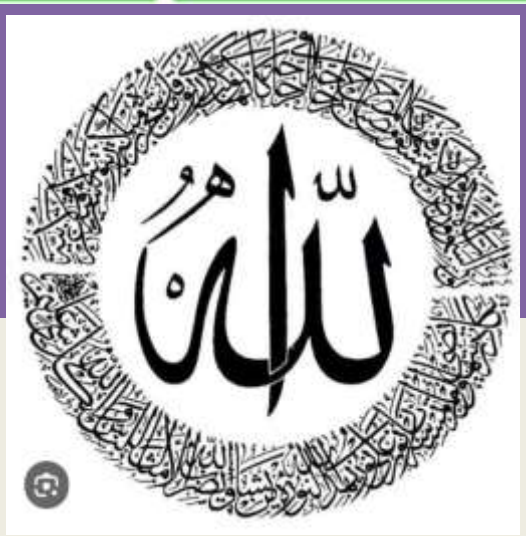
هو الله

Reverse Koebner Phenomenon in Bullous Pemphigoid – A Case Report

Abstract

Reverse Koebner response is the nonappearance or disappearance of the lesions of particular dermatoses at the site of injury. Herein we report a case of the reverse Koebner phenomenon in bullous pemphigoid in a 35-year-old male patient with sparing of the waist area which could probably be because of the phenomenon of locus majoris resistentiae. The histopathology of the lesion showed subepidermal split with a mixed inflammatory infiltrate, which was composed of predominantly eosinophils, neutrophils, and lymphocytes, but the histopathology of the spared skin showed no abnormalities. However, the direct immunofluorescence from the perilesional area

Liza Mohapatra, Kallolinee Samal, Prasenjeet Mohanty, Siddhartha Dash

Department of Skin and VD, SCB Medical College and Hospital, Cuttack, Odisha, India

which gradually increased to ound 1–3 cm. The lesions first er the trunk and gradually over of the body. The lesions used fter 5–6 days leaving behind , which were nonspreading in erosions used to get crusted in 5 days which fall off leaving -colored skin. There was no any drug intake before the of the lesion. On cutaneous few tense bullae of size

the lesion showed subepidermal split with a mixed inflammatory infiltrate, which was

Figure 1: "Bullous pemphigoid" – Tense bullae over the perineal area

How to cite this article: Mohapatra L, Samal K, Mohanty P, Dash S. Reverse koebner phenomenon in bullous pemphigoid – A case report. Indian Dermatol Online J 2019;10:692-4.

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of Merck.

Reverse isotopic response: a rarely reported phenomenon

A dermatosis sparing another unrelated previous skin disorder is a rarely encountered condition. In this communication, we present a patient with generalized contact dermatitis whose lesions spared recent papules of insect bites.

A 32-year-old woman presented with a 5-day history of a widespread skin eruption. The first lesions started on the abdomen as pruritic, red papules and were diagnosed as insect bites. Following treatment with cetirizine and mepyramine maleate plus lidocaine gel, erythema and vesiculobullous lesions developed rapidly. She was 8-weeks' pregnant, and was otherwise healthy.

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Figure 1 Widespread vesiculobullous eruption sparing discrete papular lesions

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English

Observation Letter

87(1);103-105

doi:10.25259/IJDVL_464_19

Show Sections

Clear zone phenomenon: A rare phenomenon in ichthyosis with co-existing superficial fungal infection

Mahima Agrawal, Pravesh Yadav, Jyoti Yadav, Ram Chander

Department of Dermatology and STD, Lady Hardinge Medical College and SSKH, KSCH, Delhi - 110001, India

Corresponding author: Dr. Pravesh Yadav, Department of Dermatology and STD, Lady Hardinge Medical College and

7:44 97

done from the erythematous plaque, adjacent clear zone and the margin of clear zone with ichthyotic skin. Septate fungal hyphae were demonstrated only from the erythematous plaque. Culture from the erythematous plaque was positive for *Trichophyton rubrum* while the other two areas were negative. A diagnosis of XLRI with tinea cruris was made.

Figure 1: Case 1 - Well-defined erythematous scaly plaques in the groins extending to inner thigh (red arrow), adjacent clear uninvolved zone devoid of scaling (white arrow) and generalized ichthyotic fine scales (black arrow)

Show Sections

Cookie Consent

Occurrence of Filiform Wart over Nevus Sebaceous: A Report of Two Cases of Locus Minoris Resistentiae

Sir,

Locus minoris resistentiae (LMR) is a site that offers lesser resistance or increased vulnerability than the rest of the body to the onset of disease. Trauma, irradiation, healed herpes scars, or chronic lymph stasis commonly act as LMR, leading to localization of numerous inflammatory

the scalp since birth. Her mother noticed a new growth with finger-like projections over existing old lesions since 2 months. There was a solitary well-defined verrucous hairless orange-brown plaque with filiform growth present over the scalp [Figure 1]. Biopsy from orange-brown plaque revealed orthokeratosis, acanthosis, and defective

mosaicism is rare.

Our first case, a 10-year-old female child, presented with an asymptomatic single orange-brown raised lesion over

Figure 1: Solitary well-defined verrucous hairless orange-brown plaque with filiform growth over the frontotemporal part of the scalp (case 1)

Figure 2: (
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Case Report

Warts as a common occurrence on nevus sebaceous: A case series depicting *locus minoris resistentiae*

Shuhaab Shah[1], Faizan Younus Shah[2], Uzair Khursheed Dar[3], Shah Aaqib Aslam[3]

[1]Department of Dermatology, Government Medical College, Srinagar, University of Kashmir, Jammu and Kashmir, India, [2]Lecturer, Department of Dermatology, SKIMS Medical College, Srinagar, Jammu and Kashmir, India, [3]Registrar, Department of Dermatology, SKIMS Medical College, Srinagar, Jammu and Kashmir, India

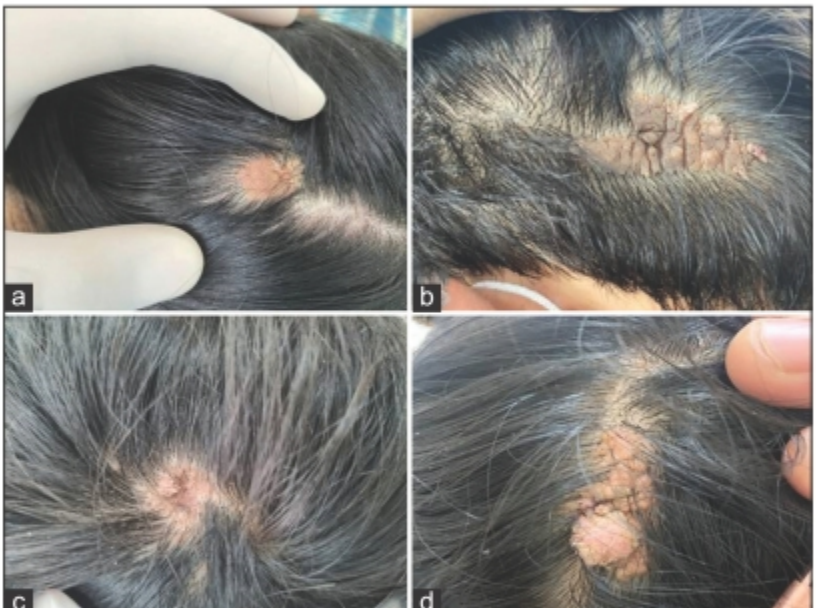

Figure 1: (a-d) Cases of nevus sebaceous with overlying warts.

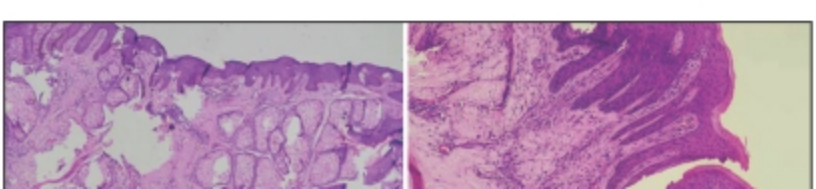

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Fig 3. **A**, Biopsy specimen shows atypical keratinocytes throughout the full thickness of the epidermis. **B**, A distinct dermal aggregate of atypical, mitotically active cells is also present. **C**, Immunostaining with pankeratin stains (AE1/AE3) highlights the epidermis. **D**, MART-1 stain highlights the dermal aggregate. (**B**, Hematoxylin-eosin stain; **C**, pankeratin stains; **D**, MART-1 stain; original magnifications: **B**, ×4; **C**, ×10; **D**, ×10.)

KEY MESSAGE

Collision tumor refers to the association of 2 or more different neoplasms within the same lesion. By virtue of their composition, they are diagnostically challenging. Dermoscopy can be a valuable tool to aid in diagnosis, allowing identification of different structures within the same lesion. Histopathologically, collision tumors show distinct neoplasms located adjacent to each other while maintaining separate boundaries.[1] Simultaneous development of 2 tumors at the same site may be coincidental; however, it is postulated that development of 1 malignancy could promote development of another via a paracrine effect or that field cancerization with or without local immunosuppression plays a role.[2] Management of collision tumors depends on the histologic features of each individual component. This patient died of metastatic melanoma.

Melanoma *in situ* and syringoma: a rare collision tumor. Clinical-pathological report of a case*

Pablo Umbert-Millet[1]
Monica Quintana-Codina[1]
Montse Salleras-Redonnet[1]

DOI: http://dx.doi.org/10.1590/abd1806-4841.20176324

Abstract: Collision or contiguous tumors, defined as two or more distinct tumors occurring at one site, are often an une
finding and may represent a diagnostic challenge, as clinical and histological presentations do not always coincide.
combinations of collision tumors have been described with respect to melanocytic lesions, with the most frequently r
being the combination of nevus and basal cell carcinoma. We present an unusual case on the nose involving a melanom
and a clinically-inapparent syringoma, which, to the best of our knowledge, is the first report of this combination.

DISCUSSION

In recent years, various terms have been used to describe two lesions occurring in one site, resulting in increasing terminological confusion. Satter *et al.* proposed a classification to simplify

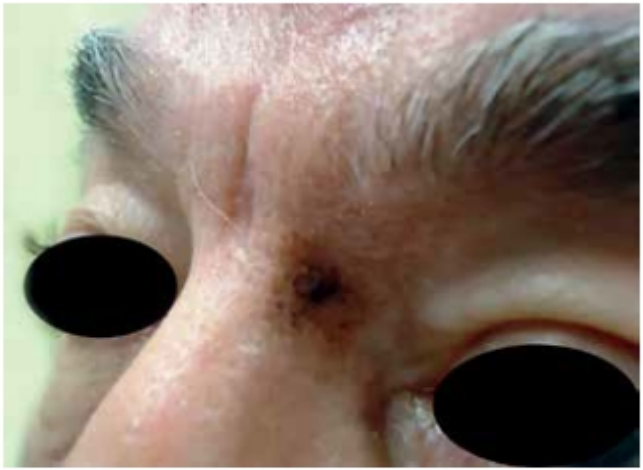
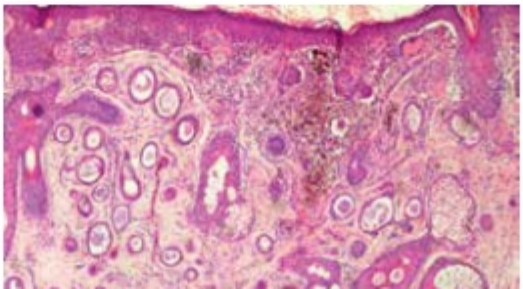


FIGURE 1: Irregular, asymmetric pigmented lesion on the left nasal root



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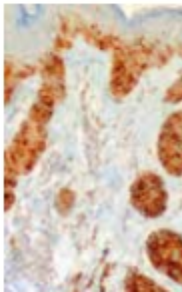

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Collision Tumor between Trichofolliculoma and Melanocytic Nevus

Christel Bolte, Roberto Cullen, Ivo Sazunic[1]

Departments of Dermatology and [1]Pathology, Clínica Las Condes, Santiago, Chile

ABSTRACT

Figure 1: A single, 5 mm diameter pink papule, with a single filiform silky hair growing through the central pore. Upon traction, the hair was 6 mm long

Figure 2: of the lesi (b) (S100 showing m Melanocyt

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CASE REPORT

A Collision Tumor: Reflectance Confocal Microscopy Features and Correlation between Dermoscopy and Histopathology

Sevil Savas, Esma Inan Yuksel[1], Aslı Vefa Erdemir[2], Mehmet Salih Gurel[2], Cem Leblebici[3]

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Abstract

Coexistence of two types of malignant neoplasms, called 'a collision tumor,' is relatively uncommon. Basal cell carcinomas (BCCs) are known to coexist with other cutaneous lesions, but the collision of BCC with malignant melanoma is rare. We report a rare case of BCC with underlying lentigo maligna melanoma, focusing on dermoscopic and reflectance confocal microscopic (RCM) findings and their correlation with histopathology. RCM and dermoscopy seem to offer important clues to increase clinical suspicion of collision tumors and improve

From the Department of Dermatology, Sultan Abdulhamid Han Research and Training Hospital, [2]Department of Dermatology, Goztepe Research and Training Hospital, Istanbul Medeniyet University, [3]Department of Pathology, Istanbul Training

erreflective nests and multiple
unding the follicular opening
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[Figure 3a and b]. The islands of
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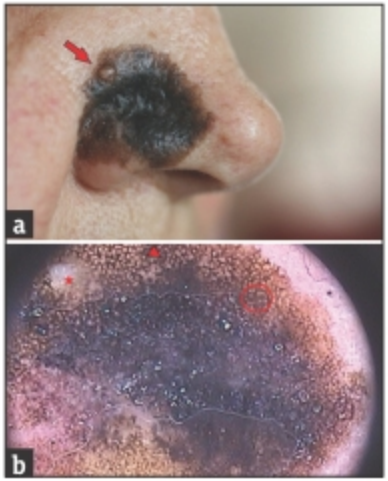

Figure 1: (a) Beige-light brown papule set on a pigmented patch (arrow). (b) Dermoscopic image shows atypical network (arrowhead), irregularly pigmented follicular openings (circle) and white homogeneous area (star)

Correlation between Dermoscopy and Histopa

Sevil Savas, Esma Inan Yuksel[1], Aslı Vefa Erdemir[2], Mehmet Salih Gurel[2], Cen

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Abstract

Coexistence of two types of malignant neoplasms, called 'a collision tumor,' is relatively uncommon. Basal cell carcinomas (BCCs) are known to coexist with other cutaneous lesions, but the collision of BCC with malignant melanoma is rare. We report a rare case of BCC with underlying lentigo maligna melanoma, focusing on dermoscopic and reflectance confocal microscopic (RCM) findings and their correlation with histopathology. RCM and dermoscopy seem to offer important clues to increase clinical suspicion of collision tumors and improve clinical diagnosis.

Key Words: *Basal cell carcinoma, collision tumor, melanoma, reflectance confocal microscopy*

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[1]Department of Derm
University, Elazıg, Tu

Address for corresp
Dr. Sevil Savas,

Kerion of the Vulva Caused by *Trichophyton mentagrophytes*

Amina Bougrine, Catherine Villeneuve-Tang, Danielle Bouffard, more... Show all authors

First Published May 1, 2014 | Case Report | Find in PubMed | Check for updates
https://doi.org/10.2310/7750.2013.13101

Article information ⌄

Altmetric 0

recorded during hospitalization was 36.6 °C.

Figure 1. Erythema and edema of the mons pubis, as well as boggy, oozing nodules involving the mons pubis, labia majora, and inguinal folds. Subtle scal[illegible]es were noted on the lower abdomen and in the inguinal folds.

↑ TOP

CASE STUDY

Abdominal Elephantiasis Nostras Verrucosa: An Underrecognized Disorder

Derek Ho, BS;[1] Eve J. Lowenstein, MD, PhD[1,2]

Case 1 A 60-year-old African American man presented with a medical history of morbid obesity (weighing more than 550 lb), benign prostatic hyperplasia, hyperlipidemia, gynecomastia, atrial fibrillation, hypertension, chronic obstructive pulmonary disease, psoriasis, sleep apnea, and cardiomegaly. His size has limited his ability to clean himself properly. He experiences chronic and recurrent skin irritation and cellulitis possibly secondary to intertriginous superinfection, lymphedema, and venous stasis. On examination, his abdomen showed heavy skinfolding with hyperpigmentation, cobblestone and velvety changes, thickening and fibrotic induration of the skin, focal areas of redness, and tenderness on palpation (Figure 1A and 1B). These findings are consistent with a clinical picture of chronic elephantiasis nostras verrucosa (ENV) and lipodermatosclerosis (LDS). He has had numerous hospitalizations for recurrent cellulitis in the abdomen and lower limbs during the past 8 years. He is currently residing in a weight-loss facility and is managing skin care using topical nystatin

250 lb), asthma, diabetes, and hyperlipidemia. On examination, he has swollen, woody lower aspects of the legs and testicular enlargement secondary to chronic lymphedema. His abdomen has pebbling, fibrosis, and thickening of the skin (Fig-

significant venous insufficiency of his legs from incompetent saphenofemoral valves. He recently started using compression stockings and doing exercises that include leg elevations and walking to enhance venous and arterial circulation.

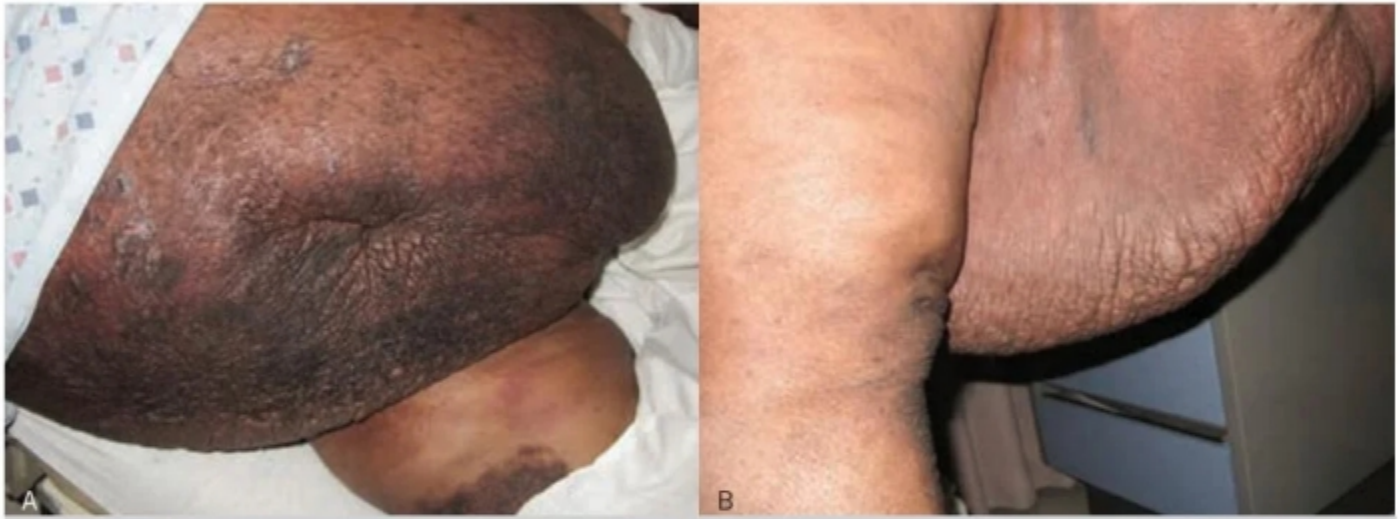

Figure 1. The patient's abdomen exhibiting hyperpigmentation and a classic elephantiasis nostras verrucosa cobblestone appearance (A). Scarring from recurrent abdominal cellulitis and focal, red nodular changes are noted (B).

From SUNY Health Science Center Department of Dermatology,[1] and Brookdale University Hospital,[2] Brooklyn, NY

Address for Correspondence: Eve J. Lowenstein, MD, PhD, SUNY Downstate Medical Center, Department of Dermatology, 8th Floor, 450 Clarkson Avenue, Box 46, Brooklyn, NY 11230 • E-mail: evlow13@yahoo.com

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Pseudomycetoma of the scalp caused by *Microsporum canis*☆,☆☆

Ligia Rangel Barboza Ruiz [a], Clarisse Zaitz [a], Rute Facchini Lellis [b], John Verrinder Veasey [a,*]

[a] *Clinic of Dermatology, Faculdade de Ciências Médicas, Santa Casa de Misericórdia de São Paulo, São Paulo,*
[b] *Pathology Laboratory, Hospital da Santa Casa de Misericórdia de São Paulo, São Paulo, SP, Brazil*

Received 28 March 2019; accepted 22 July 2019

Clinical aspect of pseudomycetoma at occipital lesion. A and B, patient with 14-years-old. C, patient with 24-years-old.

Clinical dermatology • **Concise report**

CED
Clinical and Experimental Dermatology

Blue vitiligo following intralesional injection of psoralen combined with ultraviolet B radiation therapy

J. A. Zhang, J. B. Yu, Y. Lv and P. Thapa

Department of Dermatology, First Affiliated Hospital of Zhengzhou University, Zhengzhou, Henan, China

doi:10.1111/ced.12537

Summary

A 23-year-old Chinese man presented with a 16-month history of white patches on his abdomen and neck. He had previously received an intralesional injection of psor-

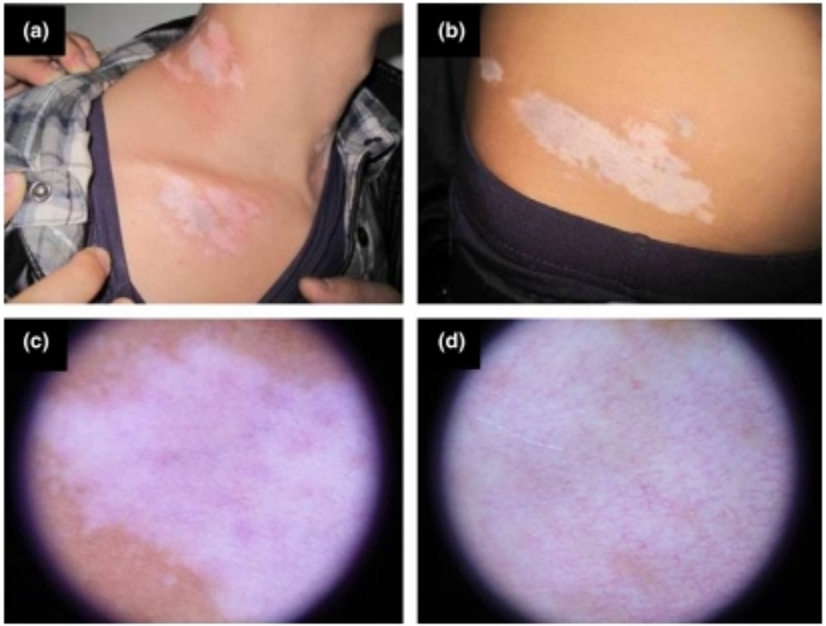

Figure 1 (a,b) Blue–grey macules over the depigmented patches on (a) neck, chest and (b) lower right abdomen. (c,d) Dermoscopy revealed blue patches and reticular telangiectasia within the white patches.

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Yellow urticaria and biliary pancreatitis

Eira Mabel Nava Cruz, MD, Juan Pablo Castanedo-Cázares, MD,
Diana Vianney Hernández Blanco, MD, and Bertha Torres-Álvarez, MD
San Luis Potosí, México

Key words: bilirubin; Hall stain; pancreatitis; yellow urticaria.

NTRODUCTION

A physical examination found large, disseminated

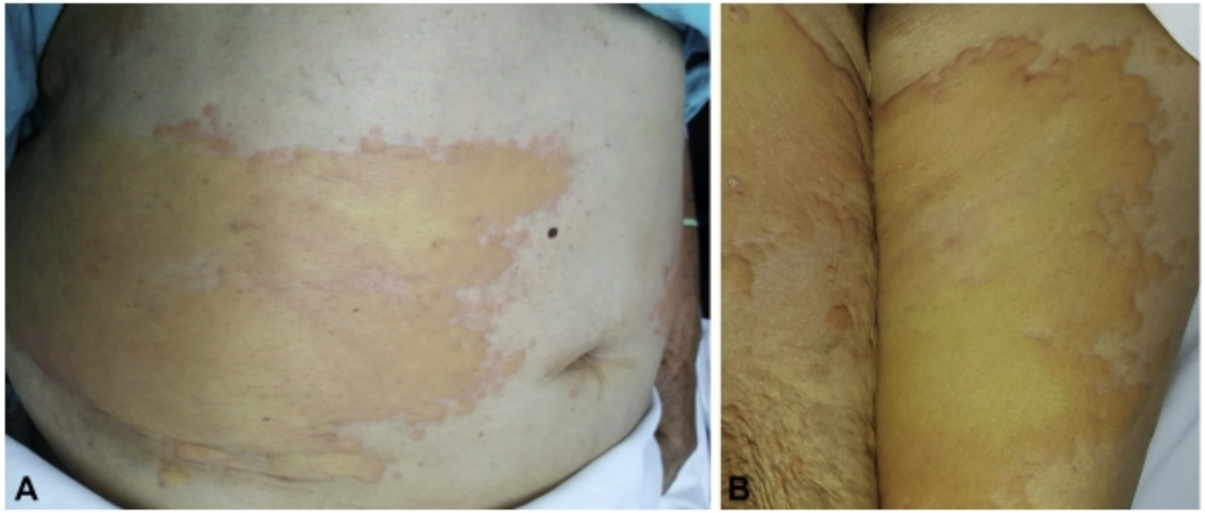

Fig 1. Yellow urticaria. **A**, Yellow raised plaque with irregular and erythematous border on the right side of the abdomen. **B**, Numerous wheals that form a large yellow plaque with a polycyclic and erythematous borders on the thighs.

Fig. 11.57 Partially hyperpigmented, sharply demarcated flat plaque of superficial basal cell carcinoma

Fig. 1.67 Shape. Polycyclic lesions are arranged in more than one ring. Note the scaling of the slightly raised margin. This is *tinea corporis*.

Tinea corporis, any questions?

op. Med. Hyg., 86(5), 2012, p. 741
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t © 2012 by The American Society of Tropical Medicine and Hygiene

Images in Clinical Tropical Medicine

Sporotrichosis Mimicking Keratoacanthoma

Fernanda Lauermann,* Marcelo Lyra, and Raquel Gaudio

Department of Dermatology, Bonsucesso Federal Hospital, Rio de Janeiro, RJ, Brazil; Evandro Chagas Clinical R
Oswaldo Cruz Foundation (FIOCRUZ), Rio de Janeiro, RJ, Brazil

2-year-old female presented with a tumor in the frontal

FIGURE 2. Tumor in the frontal region with 4 cm diameter, punctuated in its center with keratosic material.

Received November 1, 2011. Accepted for publication December 8, 2011.

Authors' addresses: Fernanda Tcatch Lauermann and Raquel Lopes Gaudio, Department of Dermatology, Bonsucesso Federal Hospital, Rio de Janeiro, RJ, Brazil, E-mails: fernandalauermann@hotmail.

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Cutaneous Chronic Graft versus Host Disease Mimicking Verruca Vulgaris

Soumya Jagadeesan, Lekshmi Sajini, Prasanna Duraisamy, Lakshmi Valliyil[1]

Department of Dermatology, Amrita Institute of Medcial Sciences, Kochi, [1]Consultant Dermatologist, Queen Haoura Aesthetic Clinic, Calicut, Kerala, India
E-mail: ammaslekshmithun@yahoo.com

Indian J Dermatol 2023:68(2):225-7

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as pityriasis-rubra-pilaris-like, ichthyosis-like, keratosis pilaris-like, psoriasiform and ezcematoid eruptions have been reported.[1]

Figure 1: Clinical images – (a) verrucous papules and plaques over the palms, (b) verrucous papules and plaques over the palms and (c) verrucous papules and plaques over the soles

Case Report - (2016) Volume 7, Issue 6

Occurrence of Necrotizing Fasciitis on Zoster Thorax in HIV Negative Patients

Aka BR[1], Diabaté A[1*], Kaloga M[2], Kourouma HS[2], Gué I[1], Ecra EJ[2], Sangaré A[2] and Yoboué YP[2]

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Figure 1: Zoster lesion.

A prescription was made including acyclovir for injection, tramadol with saline. The blood count showed mild anaemia normochromic no glucose at 0.87 g/l. The retroviral serology was negative

DOI: 10.7241/ourd.20142.59

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HERPES ZOSTER ON SEGMENTAL VITILIGO: ISOTOPIC RESPONSE?

Mankesh Lal Gambhir, Kritika Pandey, Tejinder Kaur

Department of Dermatology, Venereology & Leprosy, Government Medical College, Amritsar, Punjab, India

upport:

Figure 1. Herpes Zoster over segmental vitiligo: isotopic phenomenon.

A case of herpes simplex on scrofuloderma has been described [4,6].

Viral, immunologic, neural and vascular etiologic reasons

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Post herpes-zoster Keloid in a 10 year old boy.

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An Extraordinary Colocalization of Alopecia Areata and Vitiligo

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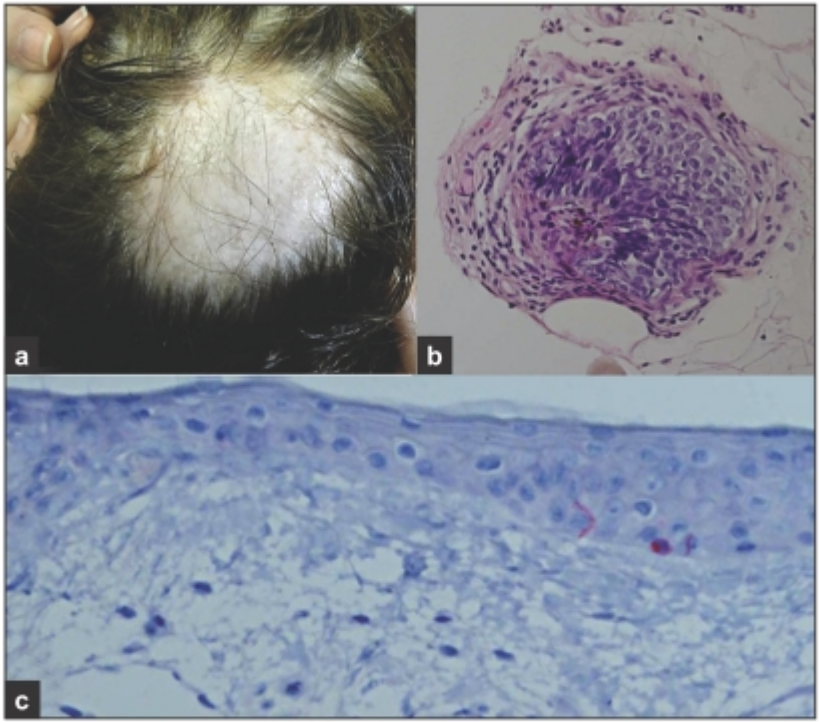

Figure 1: (a) Coexistence of loss of hair and loss of pigment in the patient's scalp; (b) Histopathology of the scalp lesion demonstrating the presence of an anagen hair follicle with a peribulbar lymphocytic infiltrate, corresponding to the diagnosis of AA; (c) MART-1 immunostaining of the same biopsy, demonstrating almost total loss of melanocytes in the basal layer, with only one or two melanocytes identified in the epidermis, verifying the coexistence of vitiligo in the same region

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UVB-induced bullous pemphigoid in a patient with psoriasis

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Bullous pemphigoid (BP) is an autoimmune subepidermal blistering disease affecting mostly elderly people. The clinical picture includes erythematous oedema, ex-

Coexistence of psoriasis and bullous pemphigoid is rare, but BP is the most frequent bullous disease described in psoriatic patients [7]. Although it is likely that

Figure 1. The patient with psoriasis and bullous pemphigoid triggered by NB-UVB treatment

Table 1. Comparison of patients' case reports with BP triggered by NB-UVB phototherapy

Patients' details	Suwarsa *et al.*	Corey *et al.*	Our patient
Age	39 years old	49 years old	71 years old
Sex	Male	Male	Male

21.00 x 29.70 cm

sciendo
Serbian Journal of Dermatology and Venereology 2018; 10 (4): 124-128

DOI: 10.2478/sjdv-2018-00

Concurrence of Bullous Pemphigoid and Psoriasis: A Case Report

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UDC 616.527:616.517

A
B

WOOD'S LAMP EXAMINATION OF THE SKIN	
Disorder/infection/colonization	**Fluorescent color/clinical findings**
Pigmentary disorders	
Vitiligo	Chalk-white to dull bluish-white (fluorescence of dermal collagen observed due to a marked decrease or absence of melanin within the epidermis)
Ash leaf spots	Enhancement of hypopigmentation
Hyperpigmentation due to an increase in:	
• epidermal melanin	Enhancement of brown color
• dermal melanin	Difference in color of lesional vs nonlesional skin becomes less obvious
Bacterial infections/colonizations	
Pseudomonas aeruginosa	Green
Corynebacterium minutissimum	Coral red
Propionibacterium acnes	Orange–red (in comedones)
Fungal infections	
Pityriasis (tinea) versicolor due to *Malassezia* spp.	Yellowish-white, yellow–green, golden, copper–orange
Tinea capitis due to *Microsporum* spp.	Blue–green to yellow–green
Favus due to *Trichophyton schoenleinii*	Blue–white

Table 0.9 Wood's lamp examination of the skin.

the dermatologist can usually determine by observation whether the

and dermatopathology both rely heavily upon careful observation and

INFECTIOUS CAUSES OF A LYMPHOCUTANEOUS ("SPOROTRICHOID") PATTERN
Most common
Atypical mycobacteria, especially *M. marinum*, but also other species (e.g. *M. chelonae*, *M. kansasii*)
Sporotrichosis (Fig. 77.24A)
Unusual
Nocardiosis (Fig. 77.24C)
Pyogenic bacteria (e.g. *Staphylococcus aureus, Streptococcus pyogenes*)
Pseudallescheria boydii/Scedosporium apiospermum
Rare (in high-income countries)
Leishmaniasis
Tularemia*
Tuberculosis*
Dimorphic fungi (other than *Sporothrix* spp.)
Opportunistic fungi in immunocompromised hosts (e.g. *Fusarium*, *Alternaria* spp.)
Glanders (*Burkholderia mallei*)*
Cat scratch disease*
Anthrax
Cowpox
Acanthamoeba spp.
*Often ulceroglandular.

Table 77.17 Infectious causes of a lymphocutaneous ("sporotrichoid")

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Resident's Page

Looking at the "Inverse" in dermatology

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ABSTRACT

The predilection of skin disorders for characteristic sites and awareness of these sites is very important for clinical diagnosis. However, certain subtypes of common dermatological conditions can involve sites opposite to that of the classical presentation. These subtypes are named using the prefix "inverse" or the suffix "inversus/inversa" in the literature. These variants may pose a diagnostic challenge to residents and knowledge of the various "inverse" dermatological conditions and phenomena in the literature is of great significance.

Keywords: Inverse, Inversus, Inversa, Inversum

INTRODUCTION

There are certain subtypes of common dermatological conditions where the distribution is opposite to the classical distribution. These subtypes are named using the prefix "inverse" or the suffix "inversus/inversa" in the literature. The Latin words "inversus" (masculine) and "inversa" (feminine) meaning "upside down" are used for these dermatological conditions. An electronic and manual search of standard dermatology textbooks and databases including PubMed/Medline and Google Scholar was done to include all the "inverse" variants of skin diseases. These uncommon to rare inverse variants add to the diverse presentation in dermatological diseases.

INVERSE PSORIASIS

This subtype (also referred to as flexural or intertriginous psoriasis) commonly involves the groins, axillae, inframammary creases, perianal, and retroauricular regions. In rare cases, it can involve the antecubital and popliteal fossae and the interdigital spaces.[1,2] The plaques of Inverse Psoriasis are thin and well-defined, with reduced or absent scaling and a glazed surface as shown in Figures 1 and 2.[3] There can be a sudden appearance of Inverse Psoriasis in HIV-infected adults.[4]

INVERSE PITYRIASIS ROSEA/PITYRIASIS ROSEA INVERSUS

When the lesions of pityriasis rosea spare the trunk and affect the axillary and inguinal folds, face, neck, and extremities, the variant is called inverse pityriasis rosea [Figure 3].[5] It is prevalent in the pediatric age group and in dark-skinned individuals.[6]

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Figure 1: Napkin psoriasis in an infant.

Figure 2: Inverse psoriasis in an adult involving axilla.

Figure 3: Inverse pityriasis rosea in a young female involving the neck.

INVERSE LICHEN PLANUS (LP)

This is an uncommon variant of LP in which the characteristic violaceous papules are seen in the axillary and inguinal folds and the inframammary areas. It can also involve other flexural areas such as the popliteal and antecubital fossae.[7] The lesions of inverse LP present as non-scaly erythematous lesions with ill-defined borders. In addition, erosions, hyperpigmentation, and hyperkeratotic papules can occur.[8]

LICHEN PLANUS PIGMENTOSUS (LPP) INVERSUS

Pock *et al.* described this rare subtype of LP in 2001.[9] LPP inversus is characterized by mildly itchy hyperchromic macules in the flexural areas, mainly the axillary folds (90%) and groins, and rarely, the antecubital and popliteal areas.[10] There is an absence of involvement in sun-exposed areas in contrast to classical LPP. The closest differential is ashy dermatosis which has a predilection for the trunk and limbs, and not the flexural areas.[11]

INVERSE GOTTRON PAPULES

Gottron's papules which are a characteristic sign of dermatomyositis are inflammatory, flat-topped, lichenoid papules on the extensor aspect of hands, particularly over the interphalangeal and metacarpophalangeal joints of the fingers.[12] When these papules are present on the palmar surfaces of the finger creases as hyperkeratotic papules, they are referred to as inverse Gottron's papules.[13] Inverse Gottron papules have been associated with severe Interstitial lung disease in adults and anti-MDA5 antibodies.[14]

ACNE INVERSA

The term "acne inversa" was proposed in 1989 by the Plewig and Steger for hidradenitis suppurativa (HS).[15] The term "acne inversa" indicates the involvement of pilosebaceous unit in the disease.[16] The histopathology and pathogenesis are similar to classical acne while the involvement of intertriginous sites is in contrast to acne. Some dermatologists are of the opinion that the term "acne inversa" should be preferred over "hidradenitis suppurativa."[17] In contrast, some are of the view that HS, though a misnomer, identifies a distinct entity that has discrete genetic basis, etiological factors, and disease associations, and therefore, the name should not be replaced.[18]

PTERYGIUM INVERSUM UNGUIS (PIU)

This is a rare condition which affects the distal part of the nail bed. The hyponychium is attached to undersurface of the nail plate. Therefore, with the growth of the nail plate, the hyponychium extends forward resulting in expungement of the distal groove.[19]

In classical pterygium unguis, the proximal nail fold fuses with the nail matrix and nail bed. PIU is otherwise called ventral pterygium. It can be congenital or acquired.[20] The acquired forms may be idiopathic or consequential to systemic conditions such as connective tissue diseases, leprosy, neurofibromatosis, or paresis.[21] Figure 4 shows pterygium inversum unguis in a patient with systemic sclerosis.

OPHIASIS INVERSUS

Muñoz and Camacho published the first description of this variant of alopecia areata in 1996. It is also known as sisaipho (ophiasis spelled backward).[22] It presents with non-scarring alopecia sparing the temporal and occipital areas [Figure 5]. Various studies have shown its increased association with atopy, vitiligo, thyroid disease, and trachyonychia.[23] The prognosis of this condition is better that of ophiasis.[23]

Figure 4: Pterygium inversum unguis in a patient of systemic sclerosis.

Figure 5: Ophiasis inversus.

INVERSE TYPE OF JUNCTIONAL EPIDERMOLYSIS BULLOSA (JEB INVERSA)

JEB inversa is a rare variant with symmetrical involvement of the body folds, that is, the axillary, inguinal, and perineal areas. The erosions heal with atrophic white scars. It has a non-lethal course with generalized blistering in the neonatal period and only flexural involvement in adult life. The proposed hypothesis is that some mutant proteins might be more unstable in the warmer regions of temperatures of the body folds.[24]

INVERSE TYPE OF RECESSIVE DYSTROPHIC EPIDERMOLYSIS BULLOSA (RDEB-I)

This is a rare variant of RDEB which presents in infancy with blistering, erosions, and atrophy over the axillae, thighs, groins, and neck. In adulthood, there is predominant involvement of the perineal area and the axillary folds.[25] Hands, feet, elbows, and knees are not involved.[26] This type of EB was first described in 1971 by Gedde Dahl.[27] It frequently involves the oral and esophageal mucosae, cornea, vulva, external ear canal, and nails. There is an increased risk of dental caries.[28] A study observed that Collagen7 from RDEB-I mutations demonstrated instability at high temperatures.[29] RDEB inversa carries a comparatively better prognosis.[27]

INVERSE KLIPPEL-TRENAUNAY SYNDROME

The classical features of Klippel-Trenaunay syndrome include capillary malformation of skin (port wine stain) with ipsilateral soft tissue and bone hypertrophy, and varicose veins. In the inverse presentation, the affected limb has deficient growth and is smaller in size. Although it has been postulated that the presence of a minus allele might be responsible for this deficient growth, the cause is still unknown.[30]

INVERSE PAPULAR ACROKERATOSIS (ACROKERATOELASTOIDES OF OSWALDO COSTA)

It is a rare genodermatosis presenting with skin-colored, horny papules on the lateral and dorsal aspects of palms and soles. It was described by Oswaldo Costa in 1952 and is seen mainly in young dark skinned women as asymptomatic keratotic papules on the lateral margins of hands and feet.[31]

There are a few more case reports of "inverse" conditions as summarized in Table 1.[32-39]

Table 2 enumerates Inverse phenomena and appearances in dermatology.[40-45]

Table 1: "Inverse" dermatological conditions mentioned in case reports.

Inverse sporotrichoid pattern of cutaneous tuberculosis[32,33]
Inverse subcorneal pustular dermatosis[34]
Inverse tinea versicolor (Tinea inversicolor)[35,36]
Inverse psoriasiform drug eruption[37]
Inverse lichenoid drug eruption[38,39]

Table 2: Inverse phenomena and appearance.

Inverse Koebner phenomenon/ Renbok phenomenon	First reported by Happle *et al*. in 1991 to describe normal hair growth seen in a psoriatic plaque in a patient with alopacia areata of the scalp.[40] The cellular alteration and change in the microenvironment by the first injury prevent the development of subsequent disease at the same site.[41]
Inverse halo phenomenon	Loss of pigmentation starts from the core of a melanocytic nevus rather than at the periphery.[42]
Inverse shouldering effect	The characteristic appearance seen in lipedema is present in females. Other names for this appearance are armchair appearance, bracelet sign, and cliff sign.[43] There is a symmetric increase in subcutaneous fat in bilateral lower limbs which stops abruptly at the level of malleoli and spares the feet.[44]
Inverse pigmentary network pattern	The reverse of the pigment network pattern on dermoscopy comprises white/lighter lines forming the grid of the network with comparatively darker areas filling the gaps. Other terms used for this pattern are negative pigment network and reversed pigment network. It is seen in melanoma, dermatofibroma, Spitz naevus, and evolving lesions of vitiligo.[45]

CONCLUSION

The involvement of sites opposite to the classical disease might pose a diagnostic challenge to a dermatology resident. Hence, knowledge of these unusual subtypes and their recognition is of utmost importance.

Ethical approval

The Institutional Review Board approval is not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The author(s) confirms that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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Toponyms in Dermatology

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Abstract

The term toponym means any name that is derived from a place name. Numerous dermatological conditions have their names derived from geographic places. Although most conditions may have some association to the place they have been derived from, some of them are fortuitous.

Key Words: *Toponyms, dermatology, places*

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The term toponym means any name that is derived from a place name. Numerous dermatological conditions have their names derived from geographic places. Although most conditions may have some association with the place they have been derived from, some of them are fortuitous. Here is a list of toponyms with explanations. Here, we have compiled a list of toponyms through the literature search in various dermatology dictionaries, textbooks and PubMed databases.

- **Acroangiodermatitis of Mali:** The term was first introduced by Mali *et al.* in 1965. Mali is also a country in West Africa. Acroangiodermatitis (synonym pseudo-Kaposi sarcoma) is an angioproliferative entity, usually related to chronic venous insufficiency or arteriovenous malformations of the legs.[1]
- **African Endemic Kaposi sarcoma:** It is a type of Kaposi sarcoma that is endemic to Africa, characterized by nodular, florid, infiltrative and lymphadenopathic types. It predominantly affects children and young adults and follows a fulminant course.[2]
- **African eye worm infection:** It is an infection caused by a nematode, loa loa and hence also called loiasis. It is prevalent in West Africa and is characterized by pruritis and migration of worms under the skin.[3]
- **African histoplasmosis:** It is an invasive fungal infection caused by Histoplasma capsulatum endemic in central and West Africa.[4]
- **African tick bite fever:** It is caused by *Rickettsiae africae*, characterized by the presence of multiple tick bites, multiple eschars, lymphadenopathy and lymphangitis.[5]
- **African trypanosomiasis:** A parasitic infection caused by *Trypanosoma brucei*, which is transmitted by tsetse fly and is characterized by the presence of painful chancre on skin, later undergoes haemolymphatic spread.[6]
- **Aleppo boil:** Aleppo is a city in Syria and this is another name for cutaneous leishmaniasis.[7]
- **American hookworm infection:** It is a parasitic infection caused by *Ancylostoma duodenale* and *Necator americanus*. Cutaneous manifestations are present during the penetration phase and invasive phase. They usually present as papulovesicles to frank bullous rash at the site of penetration with localized to generalized urticarial rash during the larva migration.[8]
- **American trypanosomiasis:** It is caused by *Trypanosoma cruzi*, and this disease is endemic to rural areas of Central and South America. Characterized by three stages, cutaneous urticarial rash is usually seen in the first stage along with fever, lymphadenopathy, painful inflammatory reaction at the site of inoculation and ulcerated nodules.[9]

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- **Andean sickness:** Related to the Andes mountain range in South America, this disease is also known as 'white leprosy' and 'valley sickness'. A skin lesion caused by cutaneous and mucocutaneous leishmaniasis presents as a skin ulcer which when cicatrizes leaves an ugly scar.[10]
- **Balsam of Peru:** Peru is a country in South America. Balsam of Peru is a resin that has been extracted from the bark of the balsam tree and used as antiseptic for minor injuries. It can also cause irritation or allergic contact dermatitis at the site of application.[11]
- **Biskra button:** Biskra is a city in Algeria, North Africa. Biskra button is a type of cutaneous leishmaniasis that is endemic to that area. Begins as a papule that enlarges to form a nodule that breaks down to form an ulcer.[12]
- **Bruton disease:** Bruton is a town present in England. However, the disease gets its name because of the mutations in the gene coding for Bruton tyrosine kinase. It is an X-linked recessive agammaglobulinemia characterized by recurrent pyogenic infections.[13]
- **Burdwan fever:** Burdwan is a city in West Bengal, India. The disease is also known as visceral leishmaniasis, endemic to this region. It is caused by *Leishmania donovani* and is characterized by earth grey color of the skin along with fever and Hepatosplenomegaly.[14]
- **Buruli ulcer:** *Mycobacterium ulcerans* causes widespread, painless cutaneous and subcutaneous necrosis. The lesion is called Buruli ulcer, after the Buruli region of Uganda.[15]
- **Calabar swelling:** Calabar is a city in Nigeria. Calabar swellings are transient pruritic subcutaneous swellings that are characteristic of Loa infection. It is prevalent in this region of West Africa.[16]
- **Calcutta ulcer:** Calcutta, now known as Kolkata, is a city in India. Calcutta ulcer is another name for cutaneous leishmaniasis.[12]
- **Congo red stain:** Congo is a country in Central Africa. This stain is used for the detection of amyloid in the skin. With this stain, the amyloid will show apple green birefringence under polarizing microscopy.[17]
- **Coast of California:** The café au lait macules of neurofibromatosis have a smooth border which is compared to the smooth borders of the Coast of California.[18]
- **Coast of Maine:** The café au lait macules of McCune Albright syndrome have a rough and irregular border which is compared to the Coast of Maine.[18]
- **Cronkhite-Canada syndrome:** This syndrome gets its name from its discoverers and not from the country. It is characterized by polyposis of gastrointestinal tract and the cutaneous triad of skin pigmentation, nail dystrophy and alopecia.[19]
- **Dead Sea regimen in psoriasis:** Dead Sea is a salt lake bordered by Jordan, Israel and The West Bank. Dead Sea water is said to have therapeutic effect on the skin in psoriasis due to its high salt content of 33% and minerals. The main elements of this climatotherapy are intensive sun exposure and bathing in the sea water.[20]
- **European blastomycosis:** Refers to cryptococcosis, which is a fungal infection caused by *Crytococcus neoformans*. Cutaneous lesions include umbilicated papules, pustules, crusted plaques, nodules and ulcers.[21]
- **Gambian trypanosomiasis:** Gambia is a country in West Africa. Gambian trypanosomiasis is endemic to this region, characterized by *Trypanosoma brucei gambiense* characterized by drowsiness, splenomegaly and cutaneous findings include an initial chancre.[22]
- **Haverhill fever:** Haverhill is a city in the United States of America. This disease is also known as rat bite fever that is characterized by intermittent fever, rash and joint pain that follows a rodent bite or ingestion of contaminated milk.[23]
- **Hibernian fever:** Hibernia is a Latin name for the island of Ireland. Hibernian fever is also known as TNF receptor-associated periodic syndrome.[24]
- **Kandahar sore:** Another name for cutaneous leishmaniasis. Kandahar is a city in Afghanistan.[12]
- **Lahore sore:** It is another name for cutaneous leishmaniasis and Lahore is a city in Pakistan.[12]
- **Madura foot:** It is a localized, chronic, suppurative and deforming granulomatous infection. It was first recognized as a disease entity in Madura (Madurai, India) and hence called Madura foot.[25]
- **Mallorca acne:** Mallorca is an island in Spain. Mallorca acne also known as acne aestivalis presents as monomorphic erythematous papules that are triggered by the hot and humid climate.[26]
- **Malta fever:** Also known as brucellosis. It is a zoonotic infection caused by *brucella species*. It can lead to various skin manifestations like papules, nodules, petechiae, purpura, ulcers, abscesses, and so on. Malta is a country in Europe to which this disease is endemic.[27]
- **Marseilles fever:** Is a rickettsial infection caused by ixodid tick endemic to the Mediterranean region. It is also known as Mediterranean spotted fever. Marseille is a city in France.[28]
- **Mediterranean familial fever:** It is a genetic autoinflammatory disorder characterized by recurrent peritonitis, pleuritis, fever, arthritis and a characteristic ankle rash.[29]
- **Morbihan disease:** Is characterized clinically by erythematous edema localized to the forehead, cheek, glabella and eyelids. It gets its name from the region Morbihan (France) from which the first patient of this disease hailed from.[30]
- **North American blastomycosis:** Is a fungal infection

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caused by *Blastomyces dermatitidis*. It starts at the site of injury as an indurated, ulcerated, chancriform lesion which is accompanied by lymphangitis and lymphadenopathy. It was once thought to be localized to North America but now is present worldwide.[31]

- **Ohio Valley Disease:** In the United States, histoplasmosis infection mainly occurs around the Ohio and Mississippi river valleys and hence is named after that region. Histoplasmosis is a granulomatous fungal infection caused by *Histoplasma capsulatum*.[32]
- **Peruvian warts:** They are red to purple nodules that are usually seen on the face and extremities seen in the cutaneous phase of Carrion disease caused by *Bartonella bacilliformis*. They are painless and prone to ulceration and bleeding. This disease is common in Peru, South America.[33]
- **Rocky Mountain spotted fever:** This disease was first discovered in the Rocky Mountain range in the United States of America. It is a tick-borne infection caused by *Rickettsia rickettsia* characterized by rash and fever.[34]
- **Rose Bengal stain:** It is a pink stain used in eyedrops to stain damaged conjunctival and corneal cells and identify damaged areas. It is named so because the pink colour of the stain resembles the forehead dot used by women from West Bengal, India.[35]
- **San Joaquin Valley fever:** Also known as coccidioidomycosis, is a benign infection caused by *Coccidioides immitis*. Rarely, it may become fatal disseminating to skin and other organs. It is said to be common in South Western America.[36]
- **South American Blastomycosis:** Also known as paracoccidioidomycosis, is a chronic granulomatous fungal disease caused by *Paracoccidioides brasiliensis*. This fungus is endemic to South America.[37]
- **Volhynia fever:** Also known as Trench fever, is caused by *Bartonella quintana* and the vector is the human body louse. Volhynia is a historic region in Central and Eastern Europe. Trench fever was said to have appeared first during the First World War. This disease observed as five-day fever among the German soldiers from the Volhynia province and hence was named so.[38]

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Conflicts of interest

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Table 1.4 Criteria for the diagnosis of Behçet's disease. International criteria for Behçet's disease – point score system: scoring ≥4 indicates Behçet's disease

Sign/symptom	Points
Ocular lesions	2
Genital aphthosis	2
Oral aphthosis	2
Skin lesions	1
Neurological manifestations	1
Vascular manifestations	1
Positive pathergy test	1*

Pathergy test is optional, and the primary scoring system does not include pathergy testing. However, where pathergy testing is conducted, one extra point may be assigned for a positive result

criteria (1987) (Table 1.5, 2014, draft of minor revision) [55], because neurological and vascular manifestations are included in minor symptoms, and the pathergy test

ANTIPHOSPHOLIPID ANTIBODY SYNDROME: SAPPORO–SYDNEY CRITERIA
At least one clinical criterion and one laboratory criterion required*
Clinical criteria
1. Vascular thrombosis • One or more objectively confirmed episodes of arterial, venous, or small vessel thrombosis occurring in any tissue or organ 2. Pregnancy morbidity • One or more unexplained deaths of morphologically normal fetuses at or after the 10th week of gestation – *or* – • One or more premature births of morphologically normal neonates at or before the 34th week of gestation because of eclampsia, pre-eclampsia, or placental insufficiency – *or* – • Three or more unexplained consecutive spontaneous abortions before the 10th week of gestation
Laboratory criteria – present on two or more occasions at least 12 weeks apart*
1. Anti-cardiolipin antibodies, IgG or IgM [>40 GPL or MPL – *or* – >99th percentile as measured by a standardized ELISA] 2. Lupus anticoagulant [detected according to the guidelines of the International Society on Thrombosis and Haemostasis] 3. Anti-β_2-glycoprotein I antibodies, IgG or IgM [>99th percentile as measured by a standardized ELISA]
*Patients who do not meet these criteria may still have antiphospholipid antibody syndrome.

Table 23.5 Antiphospholipid antibody syndrome (APLS): Sapporo–Sydney

CUTANEOUS FINDINGS IN PATIENTS WITH THE ANTIPHOSPHOLIPID ANTIBODY SYNDROME (APLS)
• Livedo reticularis, with or without retiform purpura • Cholesterol embolus-like proximal livedo reticularis with distal retiform purpura • Acral livedo reticularis • Sneddon syndrome • Livedoid vasculopathy or Degos-like lesions • Anetoderma-like lesions with microthrombosis • Raynaud phenomenon • Vasculitis-like lesions • Behçet disease-like lesions • Pyoderma gangrenosum-like ulcers • Nail fold ulcers • Splinter hemorrhages • Widespread cutaneous necrosis, typically as part of catastrophic APLS • Pseudo-Kaposi sarcoma • Superficial thrombophlebitis migrans

Table 23.6 Cutaneous findings in patients with the antiphospholipid antibody syndrome (APLS).

Fig. 23.8 Antiphospholipid antibody syndrome. A Purpura and ischemia of the distal portion of the foot. The purpuric lesions have irregular

BOX 14.4 MILLENNIUM CRITERIA FOR THE DIAGNOSIS OF ATOPIC DERMATITIS (BOS ET AL. 1998)[131]

1. Mandatory criterion
   - Presence of allergen-specific IgE
   - Historical, actual, or expected (in very young children)
   - In peripheral blood (RAST, ELISA) or in skin (intracutaneous challenge)
2. Principal criteria (2 of 3 present)
   - Typical distribution and morphology of eczema lesions: infant, childhood or adult type
   - If distribution is not typical, exclude other entity (dyshidrotic eczema, contact dermatitis, contact urticaria)
   - Pruritus
   - Chronic or chronically relapsing course

BOX 14.5 JAPANESE DERMATOLOGICAL ASSOCIATION CRITERIA (1997)[132]

An atopic diathesis:

>> Personal and/or family history of atopic diseases and the predisposition to overproduction of IgE antibodies

Must have three clinical criteria:

1. Pruritus
2. Typical morphology and distribution
3. Chronic and chronically relapsing course

the basis of true AD. In the same vein, the Japanese have created

and objective features (Table 14.1).[134–148] The large number of scoring systems available reflects the requirements of different clinical situations, including natural history and drug investigation.[149,150] Lack of comparative data precludes having a single recognized standard. Some of the scoring methods may obtain a low and positive score while the AD is severe,[151] and with most

DIAGNOSTIC CRITERIA FOR BASAL CELL NEVUS SYNDROME (BCNS)
Major criteria
1. More than two BCCs or one BCC before the age of 20 years 2. Odontogenic keratocysts of the jaw (proven by histology) 3. Three or more palmar or plantar pits 4. Bilamellar calcification of the falx cerebri 5. Bifid, fused or markedly splayed ribs 6. First-degree relative with BCNS
Minor criteria
1. Macrocephaly (determined after adjustment for height) 2. Congenital malformations: cleft lip or palate; frontal bossing; "coarse face"; moderate or severe hypertelorism 3. Other skeletal abnormalities: Sprengel deformity*; marked pectus deformity; marked syndactyly of the digits 4. Radiographic abnormalities: bridging of the sella turcica; vertebral anomalies such as hemivertebrae and fusion or elongation of the vertebral bodies; modeling defects of the hands and feet; flame-shaped lucencies of the hands or feet 5. Bilateral ovarian fibromas 6. Medulloblastoma
*Unilateral elevation of smaller-sized scapula.

Table 108.3 Diagnostic criteria for basal cell nevus syndrome (BCNS). The diagnosis of BCNS requires *two major or one major and two minor criteria*. BCC, basal cell carcinoma. *Adapted from ref. 50.*

Table 1 Diagnostic criteria for nevoid basal cell carcinoma syndrome (adapted from [23])

Major criteria

1. Basal cell carcinoma before 20 years of age or excessive numbers of basal cell carcinomas out of proportion to prior sun exposure and skin type
2. Keratocystic odontogenic tumor before 20 years of age
3. Palmar or plantar pitting
4. Lamellar calcification of the falx cerebri
5. Medulloblastoma, typically desmoplastic
6. First degree relative with nevoid basal cell carcinoma syndrome

Minor criteria

1. Rib abnormalities
2. Other specific skeletal malformations and radiologic changes (i.e., vertebral anomalies, kyphoscoliosis, short fourth metacarpals, postaxial polydactyly)
3. Macrocephaly
4. Cleft lip or palate
5. Ovarian or cardiac fibroma
6. Lymphomesenteric cysts
7. Ocular abnormalities (i.e., strabismus, hypertelorism, congenital cataracts, glaucoma, coloboma)

Table 2 Differential diagnosis for multiple basal cell carcinomas occurring at an early age

Table 7.4

Diagnostic criteria for NBCCS

Diagnosis based on two major or one major and two minor criteria.

Major criteria

1. More than 2 BCCs or one under age of 20 yrs
2. Odontogenic keratocyst
3. Three or more palmar pits
4. Bilamellar calcification of falx cerebri
5. Bifid, fused or splayed ribs
6. First degree relative with NBCCS

Minor criteria

1. Macrocephaly adjusted for height
2. Frontal bossing, cleft lip/palate, hypertelorism
3. Sprengel deformity, pectus, syndactyly of digits
4. Bridging of sella turcica, hemivertebrae, flame-shaped radiolucencies
5. Ovarian fibroma
6. Medulloblastoma

Based on V.E. Kimonis et al {1273}.

TABLE 1 Diagnostic Criteria of NBCCS Taken From Kimonis et al.

Major Criteria	*Minor Criteria*
1. More than two BCCs or one BCC in patients younger than 20 years of age	1. Macrocephaly
2. KCOTs of the jaw (proven by histologic analysis)	2. Congenital malformations (e.g., CL/P, frontal bossing, coarse faces, and moderate or severe hypertelorism)
3. Three or more palmar or plantar pits	3. Other skeletal abnormalities (e.g., sprengel deformity, marked pectus deformity, and marked syndactyly of the digits)
4. Bilamellar calcification of the falx cerebri	4. Radiological abnormalities (e.g., bridging of the sella turcica, vertebral anomalies, modeling defects of the hands and feet, or flame-shaped lucencies of the hands and the feet)
5. Bifid, fused, or markedly splayed ribs	5. Ovarian fibroma or medulloblastoma
6. A first-degree relative with NBCCS	

If the patient meets at least 2 major or 1 major and 2 minor criteria, it can be diagnosed as NBCCS.

Table 5 Summary of clinical features and diagnostic criteria for nevoid BCC syndrome (2 major criteria or 1 major and 2 minor criteria are required for the diagnosis) (El Safoury et al. 2013; Salem et al. 2013; Goldsmith et al. 2012b; Spitz and Hatch 2005a)

Major criteria
BCC prior to 20 years old or excessive number of BCCs
Odontogenic keratocysts of the jaw
Palmoplantar pitting (>3)
Lamellar calcification of the falx cerebri
Bifid, fused ribs
First-degree relation to nevoid BCC syndrome
PTCH mutation on normal tissue
Minor criteria
Macrocephaly
Orofacial abnormalities including cleft lip and palate, frontal bossing, hypertelorism, and coarse face
Other skeletal abnormalities including marked syndactyly, pectus deformity, Sprengel deformity, and falx cerebri calcification
Medulloblastoma
Radiologic abnormalities including bridging of sella turcica, vertebral anomalies, and flame- shaped lucencies of hands and feet
Ovarian fibroma

Table 1.3 Criteria for the diagnosis of Behçet's disease. Criteria for the diagnosis of Behçet's disease by International Study Group [63]

Findings	Definition
Recurrent oral ulcerations	Minor aphthous, major aphthous, or herpetiform ulceration observed by physician or patient, which recurred at least 3 times in one 12-month period
Plus 2 of the following	
Recurrent genital ulceration	Aphthous ulceration or scarring observed by physician or patient
Eye lesion	Anterior uveitis, posterior uveitis, or cells in vitreous on slit lamp examination or retinal vasculitis observed by ophthalmologist
Skin lesions	Erythema nodosum observed by physician or patient, pseudofolliculitis or papulopustular lesions, or acneiform nodules observed by physician in postadolescent patients not on corticosteroids
Positive pathergy test	Test interpreted as positive by the physician at 24–48 h

Note: Findings are applicable if no other clinical explanation is present

country, though there are some regional differences in other symptoms. For example, gastrointestinal involvement is more common in Far Eastern Asia such as Japan,

Table 11.2 International classifications criteria for Behçet disease – pediatric criteria for BS [14]

Item	Description	Value/ item
Recurrent oral aphthosis	At least 3 attacks/year	1
Genital ulceration	Typically with scar	1
Skin involvement	Necrotic folliculitis, acneiform lesions, erythema nodosum	1
Ocular involvement	Anterior or posterior uveitis, retinal vasculitis	1
Neurological signs	With the exception of isolated headaches	1
Vascular signs	Venous thrombosis, arterial thrombosis, arterial aneurysm	1

is classified with BS if he has a score of 3 or more (Table 11.2). These criteria are provisional and require external validation.

Unusual presentation of psoriasis

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ABSTRACT

Figure 1: Note the whorled pattern of psoriatic plaques.

CASE REPORT

Multiple linear eccrine spiradenomas with eyelid involvement

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ABSTRACT

fig. 1 Multiple eccrine spiradenomas on the face arranged in a linear configuration.

25 january 2026

(1) Nail diseases

1. Absent lunulae =Anolunula
2. Acro osteolysis
3. Acropathie ulcéro mutilante / Acropathia ulcero-mutilans
4. Alopecia areata: nail signs
5. Amelanotic melanoma of nail
6. Aneurysmal bone cyst
7. Angioleiomyoma
8. Anonychia
9. Anonychia–onychodystrophy with brachydactyly type b and ectrodactyly
10. Anonychia with bizarre flexural pigmentation
11. Asymmetric gait nail unit syndrome
12. Basal cell carcinoma
13. Beau's lines
14. Bidet nail
15. Bifid thumb
16. Blue /Azure lunulae
17. Bowen disease of nail
18. Botryomycoma
19. Brachyonychia (short nails)
20. Brittle nail
21. Brittle nail syndrome
22. Bullous diseases :nail signs
23. Carpal tunnel syndrome
24. Chevron nail
25. Chromonychia
26. Circumferential nail
27. Cirsoid angioma
28. Claw-like nail
29. Clubbing of nails

30. Coccal nail-fold angiomatosis
31. Congenital hypertrophy of the lateral fold of the hallux
32. Crumbling of nail
33. Curly fingernails
34. Curved nail of the fourth toe
35. Darier disease: nail signs
36. Dermatofibrosarcoma protuberans of nail
37. Dermatomyositis nail signs
38. Distal nail embedding
39. Distal subungual keratosis
40. Dolichonychia (long nails)
41. Double nail anomaly
42. Drug-Induced Nail Abnormalities
43. Dyskeratosis congenita: nail
44. Dystelephalangy= Kirner deformity
45. Ectopic nail =onychoheterotopia
46. Ectrodactyly
47. Eczema : nail signs
48. Elkonyxis
49. Erythronychia
50. Familial nail dysplasia
51. Familial nail dystrophy
52. Familial twenty-nail dystrophy
53. Fibrokeratoma of nail
54. Fibroma
55. Fibromyxoma
56. Subungual filamentous tumor
57. Finger sucking
58. Frictional melanonychia
59. Glomus tumour
60. Graft vs host disease :nail signs
61. Granular cell tumor
62. Grasp reflex multiple ingrown fingernails

63. Green nail syndrome =Goldman-Fox syndrome
64. Habit tic deformity
65. Half and half nail
66. Hallux
67. Hangnail
68. Haplonychia
69. Harpoon nail
70. Heloma
71. Herpes simplex :nail
72. Hooked nail
73. Hyperonychia
74. Hyponychia
75. Hypoplasia
76. Idiopathic polydactylous longitudinal erythronychia
77. Ingrown toe nail= onychocryptosis
78. involuted toenai
79. Iso kikuchi=congenital onychodysplasia of index finger COIF
80. Kaposi sarcoma
81. Keratoacanthoma
82. Keratin granulations
83. Koilonychia (spoon nail)
84. Koenen's periungual fibromas
85. Lacquer nail
86. Leprosy nail signs
87. True leukonychia: punctate & striate & diffuse leukonychia (porcelain nails & leukonychia totalis)
88. Leukonychia variegata
89. Apparent leukonychia
90. Leukonychia totalis with epiphyseal dysplasia (Lowry–Wood) syndrome
91. Leukonychia totalis with PPK and congenital alopecia
92. Leukonychia with pili torti

93. Leukonychia, axonal neuropathy, myopathic features, dilated cardiomyopathy, conduction disturbances and arrhythmia
94. Leukonychia, duodenal ulcer, and gallstones
95. Leukonychia, onychorrhexis, hypoparathyroidism, dental changes, cataract syndrome
96. Leukonychia with koilonychias
97. Striated leukonychia with eruptive milia
98. Lichen planus : nail signs
99. Lipoma
100. Longitudinal furrows
101. Longitudinal grooves
102. Longitudinal ridges
103. Macronychia
104. Malalignment of the great toenails
105. Median canaliform dystrophy of heller
106. Mee's line
107. Megalodactyly
108. Melanonychia
109. Microlunula
110. Micronychia
111. Micronychia with trisomy 21
112. Muehrcke's line
113. Myxoid cyst= Myxoid pseudocysts of the digits
114. Nail apparatus melanocytic naevus
115. Nail apparatus melanoma
116. nail degloving syndrome
117. Nail apparatus lentigo
118. Nail beads
119. Nail biting
120. Nail degloving syndrome
121. Nail dystrophy
122. Nail dystrophy–deafness syndrome= robinson's syndrome
123. Nail fragility

124. Nail fragility syndrome
125. Nail patella syndrome = Hereditary Onycho-Osteodysplasia= Fong Disease
126. Nail ridges
127. Neapolitan nail
128. Oblique lines=chevron nail=herringbone nail
129. Oil spot
130. Onychauxis
131. Onychoatrophy
132. Onychoblastoma
133. Onychocryptosis
134. Onychocytic carcinoma
135. Onychocytic matricoma
136. Onychodaknomania
137. Onychodystrophy
138. Onychogryphosis= ram's horn nail
139. Onychoheterotopia
140. Onycholemmal carcinoma
141. Onycholemmal cysts
142. Onycholemmal horn
143. Onycholysis = plummer's nails
144. Onychomadesis
145. Onychomatricoma
146. Onychomycosis (distal and lateral subungual & superficial white & proximal subungual & mixed pattern & total dystrophic & endonyx)
147. Candida onychomycosis
148. Onychopapilloma
149. Onychophagia= nail biting
150. Onychophosis =onychoclavus
151. Onychorehexis
152. Onychoschizia =lamellar splitting
153. Onychoteiromania

154. Onychotemnomania
155. Onychotillomania
156. Osteochondroma
157. Partial hereditary onycholysis
158. Acute Paronychia
159. Chronic Paronychia
160. Pachyonychia congenita
161. Parakertaotic/parakeratosis pustulosa (hjorth-sabouraud syndrome)
162. Parrot-beak nails
163. photo-onycholysis
164. Pincer nail=trumpet nails
165. Pityriasis rubra pilaris :nail signs
166. Pitting
167. Platonychia
168. Plicated nail
169. Poroma
170. Post-traumatic ectopic nail
171. Pseudomycotic nail dystrophy
172. Psoriasis :nail signs
173. Pterygium
174. Pyogenic granuloma
175. Racquet nails (nail en raquette)
176. Ragged cuticles
177. Red lunulae
178. Retronychia
179. Rheumatoid arthritis :nail signs
180. Rippled nail
181. Round fingerpad
182. Rubronychia
183. Scabies: nail signs
184. Schwannoma
185. Scleromyxedema

186.Seborrheic keratosis
187.Shell nail syndrome
188.Splinter heamorhage
189.Splitting
190.Squamous cell carcinoma (SCC)
191.Stippled nails
192.Subungual calcification
193.Subungual exostosis
194.Subungual heamatoma
195.Subungual heloma = corn
196.Subungual hyperkeratosis
197.Subungual melanoma
198.Subungual osteochondroma
199.Subungual seborrheic keratosis
200.Systemic lupus: nail signs
201.Systemic sclerosis: nail signs
202.Terry nail
203. Tile nail
204.Trachyonychia = Twenty nail syndrome
205.Transverse overcurvature of the nail
206.Trapezoidal nail
207.Triangular lunula
208.Tulip fingers
209.Upslanting toenail
210.Upward growth of nail
211.Wart
212.Worn down
213.Xanthogranuloma of nail
214.Xantholeukonychia
215.Xanthonychia
216.Yellow Lunula
217.Yellow nail syndrome

(2) Diseases of skin and mucous membranes

1. **Aarskog syndrome= facial digital genital syndrome**
2. **ABCD syndrome= black locks- albinism-deafness**
3. **Abcess**
4. **Ablution dermatitis**
5. **Abrikossoff's tumour**
6. **Abuse (Child& partner &elder&sexual)**
7. **Acatalasemia**
8. **Acanthamebiasis**
9. **Acanthoma/granuloma fissuratum**
10. **Acanthosis nigricans**
11. **Nevoid type Acanthosis nigricans**
12. **Acanthosis nigricans-like mycosis fungoides**
13. **Acantholytic acanthoma**
14. **Acantholytic dyskeratotic acanthoma**
15. **Accessory digit= Supernumerary digit=Rudimentary Polydactyly**
16. **Accessory ear pinna**
17. **Accessory nipple= supernumerary nipples**
18. **Accessory oral cavity**
19. **Accessory tragus**
20. **Achenbach syndrome=Paroxysmal hand hematoma**
21. **Achondroplasia =short limbed dwarfism**
22. **Aciduria**
23. **Acinic cell carcinoma**
24. **Acne aestivalis/ estival**
25. **Aquagenic acne**
26. **Acne chlorance**
27. **Acne conglobata**
28. **Acne cosmetica**
29. **Acne detergent**
30. **Acne excoriee=acne excoriée des jeunes filles**
31. **Acne-free' naevus**

32. **Acne fulminans**
33. **Acne fulminans sine fulminans**
34. **Acne halogens**
35. **Acne hemorrhagica**
36. **Hormonal acne**
37. **Infantile acne**
38. **Acne keloidalis nuchea= folliculitis keloidalis nuchae= folliculitis scleroticans nuchae= Dermatitis papillaris capillitii**
39. **Maskne**
40. **Acne mallorca**
41. **Acne mcdonald**
42. **Acne mechanicha & fiddler's neck**
43. **Mid-childhood acne**
44. **Acne necrotica miliaris**
45. **Acne necrotica varioliformis**
46. **Acne neonatal**
47. **Munro acne nevus**
48. **Oil acne**
49. **Acne penile**
50. **Acne pitch**
51. **Acne pomade**
52. **Preadolescent acne**
53. **post adolescent acne**
54. **Postmenopausal Acne**
55. **Acne scar (ice pick&rolling&boxcar&pitted&stellate& crateriform scars& post acne erythema &keloid &lipoatrophic& papular&hypopigmented)**
56. **Senile Acne = favre – Racouchot syndrome=solar comedones**
57. **smoker Acne**
58. **Tar acne**
59. **Acne toilet**
60. **tropical Acne**
61. **Acne rosacea**
62. **Acne due to radiation = radiation-induced comedones**

63. **Acne vasculitis**
64. **Acne venenata**
65. **Acne vulgaris**
66. **Acneform drug eruption**
67. **Steroid induced acne**
68. **Acnitis =acne agminata**
69. **Acquired bilateral melanosis of the neck in the perimenopausal woman (ABMNPW)**
70. **Acquired bilateral nevus of ota- like macules (ABNOM)**
71. **Acquired bilateral telangiectatic macules (ABTM)**
72. **Acquired blaschkoid dermatitis**
73. **Acquired Dermal Macular Hyperpigmentation**
74. **Acquired dermal melanocytosis**
75. **Acquired dyskeratotic leukoplakia**
76. **Acquired elastotic hemangioma**
77. **Acquired facial blaschkoid melanocytic nevus**
78. **Acquired focal dermal elastosis**
79. **Acquired idiopathic generalized anhidrosis**
80. **Acquired immunodeficiency syndrome (AIDS)– HIV**
81. **Acquired linear hyperpigmentation of the lips (ALHL)**
82. **Acquired Palmoplantar keratoderma**
83. **Acquired perforating dermatosis =kyrle disease**
84. **Acquired progressive lymphangioma**
85. **Acquired progressive kinking of the hair=symmetrical circumscribed allotrichia**
86. **Acquired reactive digital fibroma**
87. **Acquired symmetrical acrokeratoderma**
88. **Acquired universal melanosis = carbon baby syndrome**
89. **Acral angioosteoma cutis**
90. **Acral arteriolar ectasia**
91. **Acral fibromyxoma**
92. **Acral histiocytic nodules**
93. **Acral keratosis with eosinophilic dermal deposits**

94. **Acral peeling skin syndrome**
95. **Acral pseudolymphomatous angiokeratoma APACHE**
96. **Acral psoriasiform hemispherical papulosis**
97. **Acral speckled hypomelanosis (ASH)**
98. **Acroangiodermatitis of mali= pseudo-kaposi sarcoma**
99. **Acroangiodermatitis of stewart bluefarb syndrome (SBS)**
100. **Acrocephalosyndactyly**
101. **Acrocyanosis**
102. **Acrodermatitis acidemia**
103. **Acrodermatitis chronica atrophicans of lyme**
104. **Acrodermatitis continua of hallopeau=dermatitis repens**
105. **Acrodermatitis dysmetabolica**
106. **Acrodermatitis enteropathica**
107. **Acrodermatitis pustulosa**
108. **Acarodermatitis urticarioides**
109. **Acrodynia=epidemic erythema =calomel disease = pink disease=erythredema polyneuropathy**
110. **Acrodysostosis**
111. **Acrogeria=Gottron syndrome**
112. **Acrokeratoelastoidosis of oswald costa**
113. **Acrokeratosis paraneoplastica =Bazex syndrome**
114. **Acrokeratosis verruciformis of hopf**
115. **Acromegaloid phenotype with cutis verticis gyrata and corneal leukoma**
116. **Acromelanosis**
117. **Acromelanosis albo-punctata**
118. **Achromia parasitica**
119. **Acroosteolysis**
120. **Acroosteolysis with osteoporosis= Hajdu–Cheney syndrome**
121. **Acropathia ulcero-mutilans acquisita=Bureau–Barrière Thomas syndrome**
122. **Acropustulosis of infancy**
123. **Acrospiroma**
124. **Actinic comedonal plaque**
125. **Actinic granuloma o' brien**

126. **Actinic /senile/solar keratosis: (atrophic & actinic cheilitis & hypertrophic (hyperkeratotic) & lichenoid & pigmented)**
127. **Chronic actinic dermatitis CAC = photosensitivity dermatitis= photosensitive eczema**
128. **Actinic reticuloid**
129. **Actinic prurigo= hutchinson's summer prurigo =familial hereditary polymorphous light eruption of american Indians**
130. **Acinetobacter-baumannii-associated Infection**
131. **Actinomycosis**
132. **Actinobacillosis**
133. **Activated pi3kδ syndrome**
134. **Acute bullous hemorrhagic prurigo**
135. **Acute cutaneous necrosis syndrome**
136. **Acute digital gangrene in newborn**
137. **Acute generalized exanthematous pustulosis AGEP = toxic pustuloderma**
138. **Acute hemorrhagic edema of infancy AHEI= finkelstein–seidlmayer disease=Cockarde Purpura and Edema=Postinfectious cockade purpura**
139. **Acute inflammatory edema (AIE)**
140. **Acute syndrome of apoptotic pan epidermolysis**
141. **Adam17 deletion=neonatal-onset inflammatory skin and bowel disease**
142. **Adams oliver syndrome=Congenital scalp defects with distal limb anomalies**
143. **Adducted thumbs syndrome**
144. **Adeno virus related diseases**
145. **Adenocarcinoma**
146. **Adenoid cystic carcinoma**
147. **Adenolipoma**
148. **Adenomas in minor vestibular glands**
149. **Adenosine deaminase 2 (ADA) deficiency**
150. **Adenosquamous carcinoma**
151. **Adermatoglyphia**
152. **Adnexal adenocarcinoma not otherwise specified AANOS**
153. **Adnexal polyp of neonatal skin**

154. **Adrenoleukodystrophy= schilder's disease**
155. **Adult gluteal granuloma=granuloma gluteale adultorum**
156. **Adult-onset still disease (AOSD)**
157. **AESOP syndrome : adenopathy, extensive skin patch overlying plasmacytoma**
158. **African tick bite fever**
159. **Aggressive digital papillary adenocarcinoma**
160. **Aggressive digital adenoma**
161. **Aicardi–goutières syndrome**
162. **Ainhum**
163. **Aggressive digital papillary adenoma**
164. **Alagille syndrome**
165. **Albinism & oculocutaneous albinism (7types)**
166. **Albright hereditary osteodystrophy**
167. **Aleukemic leukemia cutis**
168. **Alezzandrini syndrome**
169. **Algal infection**
170. **Alkaptonuria**
171. **Allgrove's syndrome**
172. **Alopecia-contractures-dwarfism mental retardation syndrome**
173. **Ant- induced alopecia**
174. **Accidental alopecia**
175. **Androgenetic alopecia AGA =common baldness=male or female pattern hair loss**
176. **Diffuse patterned alopecia (DPA)**
177. **Diffuse unpatterned alopecia (DUPA)**
178. **Reverse pattern androgenetic alopecia**
179. **Alopecia areata =area celsi (patchy&totalis &universalis &ophiasis & linear& perinevoid)**
180. **Alopecia areta ophiasis inversa= sisaipho**
181. **Alopecia areata incognito**
182. **Central centrifugal cicatricial alopecia = hot comb alopecia**
183. **Chemotherapy-induced alopecia**

184. Cicatricial /scarring alopecia
185. Congenital Triangular Alopecia CTA
186. Alopecia of DLE discoid lupus erythematosus
187. Fibrosing alopecia in a pattern distribution (FAPD)
188. Frontal fibrosing alopecia FFA
189. Alopecia mucinosa
190. Alopecia neoplastica
191. Noncicatricial alopecia from systemic lupus erythematosus
192. Occipital Alopecia
193. Alopecia parvimaculata
194. Premature alopecia
195. pseudopelade of brocq= Alopecia Atrophicans= Alopecia Cicatrisata
196. Pressure induced alopecia=post-operative alopecia
197. Psoriatic alopecia
198. Radiation-induced alopecia
199. Retrograde alopecia
200. Senescent alopecia
201. Alopecia syphilitica
202. Toxic alopecia
203. Tractional alopecia
204. Traumatic alopecia
205. Congenital Alopecia universalis
206. Varicella-zoster virus (VZV) associated alopecia
207. Alopecic and aseptic nodule of scalp
208. Alopecia-onycho-dysplasia-hypohidrosis-deafness Syndrome
209. Alopecia–anosmia–deafness–hypogonadism (johnson neuroectodermal syndrome)
210. Anterolateral alopecia of the legs= peroneal alopecia
211. Alport syndrome
212. Alpha-gal syndrome
213. Alteranariosis
214. Alveolar soft part sarcoma
215. Amalgam tooth related diseases

216. **Amalgam tattoo**
217. **Amaurosis congenita, cone–rod type, with congenital hypertrichosis**
218. **Amastia**
219. **Amazia**
220. **Amoebiasis cutis**
221. **Ameloblastoma**
222. **Amelo-onychohypohidrotic dysplasia**
223. **Amelo-cerebrohypohidrotic syndrome (kohlschutter–tonz syndrome; epilepsy and yellow teeth syndrome)**
224. **American tick bite fever=Maculatum disease**
225. **Amicrobial pustulosis of fold**
226. **Amish brittle hair syndrome(BIDS):Brittle Hair&Intellectual Impairment&Decreased Fertility&Short Stature&Nail Dystrophy)**
227. **Amniotic constriction band (amnion rupture sequence)**
228. **Amputee-related dermatoses**
229. **Amyloidosis (see next)**
230. ✓ **Systemic**
231. ✓ **Dialysis related amyloidosis**
232. ✓ **Amyloidosis cutis dyschromia**
233. ✓ **Macular**
234. ✓ **Familial amyloidosis, finnish type**
235. ✓ **Familial cutaneous amyloidosis**
236. ✓ **Frictional**
237. ✓ **Biphasic**
238. ✓ **Nodular**
239. ✓ **Lichen**
240. ✓ **Bullous**
241. ✓ **Anosacral**
242. ✓ **Atrophic nodular cutaneous amyloidosis (ANCA)**
243. ✓ **Poikiloderma-like cutaneous amyloidosis**
244. ✓ **Familial amyloidotic polyneuropathy**

245. ✓ **Primary amyloidosis of the external ear (PAEE)=Auricular collagen papules**
246. ✓ **Secondary**
247. **Amyloid elastosis**
248. **Anagen effluvium**
249. **Anasarca-induced desquamation**
250. **Ancylostomiasis**
251. **Anetoderma =Atrophia maculosa cutis (Primary: jadassohn–pellizzari & schweninger-buzzi anetoderma & secondary & of prematurity &familial)**
252. **Angelman syndrome**
253. **Angina bullous heamorhagica= benign hemorrhagic bullous stomatitis**
254. **Angioedema**
255. **Angiofibroma**
256. **Angiohistiocytoma**
257. **Angioinvasive fungal infections**
258. **Angiokeratoma (solitary &circumscriptum & mibelli & corporis diffusum& fordyce)**
259. **Angioleiomyoma**
260. **Angiolipoma**
261. **Angiolipoleiomyoma**
262. **Angiolymphoid hyperplasia with eosinophilia (ALHE)= epithelioid hemangioma=pseudopyogenic granuloma=popular angioplasia =histiocytoid hemangioma**
263. **Angioma serpignosa= progressive patchy capillary malformations**
264. **Angiomatosis**
265. **Angiomatoid fibrous histiocytoma=angiomatoid malignant fibrous histiocytoma**
266. **Glomeruloid reactive angioendotheliomatosis**
267. **Malignant angioendotheliomatosis**
268. **Reactive angioendotheliomatosis (RAE)**
269. **Angiomyofibroblastoma**
270. **Angiomyolipoma**
271. **Angiomyomatous hamartoma**

272. **Superficial Angiomyxoma**
273. **Deep Angiomyxoma**
274. **Angiopericytomatosis**
275. **Angiosarcoma=lymphangiosarcoma=hemangiosarcoma**
276. **Angora hair nevus Syndrome =Schauder syndrome**
277. **Angry back syndrome**
278. **Anhidrosis**

Animals in dermatology (see next)

279. ✓**Alligator boy**
280. ✓**Bird-like facies**
281. ✓**Bovine popular stomatitis**
282. ✓**Buffalo hump**
283. ✓**Bulldog jaw**
284. ✓**Bull's eye appearance**
285. ✓**Butterfly disease**
286. ✓**Butterfly effect**
287. ✓**Butterfly pox**
288. ✓**Butterfly rash**
289. ✓**Butterfly sign**
290. ✓**Cat bite sign**
291. ✓**Cat scratch disease**
292. ✓**Cat tongue sign**
293. ✓**Caterpillar bodies**
294. ✓**Caterpillar dermatitis**
295. ✓**Caviar appearance on dermoscopy of ochronosis**
296. ✓**Caviar tongue**
297. ✓**Chicken skin/Moroccan leather appearance**
298. ✓**Chickenpox**
299. ✓**Chicken wire appearance/pattern**
300. ✓**Chicken wire erythema**
301. ✓**Cowboy gunbelt and holster**

302. ✓Crab louse
303. ✓Crab yaws
304. ✓Crow's feet
305. ✓Crowe's sign
306. ✓Dog-faced hypertrichosis lanuginose
307. ✓Eggshell nails
308. ✓Elephantiasis neurofibromatosa
309. ✓Elephantine psoriasis
310. ✓Elephantiasis nostras verrucosa
311. ✓Elephantiasic variant thyroid dermopathy
312. ✓faun tail nevus
313. ✓Fish like scales
314. ✓Fishman syndrome
315. ✓Fish mouth wounds
316. ✓Fishnet-like pattern
317. ✓Fish odor syndrome
318. ✓Fish pedicure
319. ✓Fish skin graft
320. ✓Fish tail deformity
321. ✓Fish tank granuloma
322. ✓Fox–Fordyce disease
323. ✓Fried egg appearance
324. ✓Frog-spawn appearance
325. ✓Goose skin
326. ✓Horse rider's pernio
327. ✓Horseshoe-shaped' rim of hair
328. ✓Horse-riding stance
329. ✓Horse-collar pattern
330. ✓LAMB syndrome
331. ✓Leopard skin
332. ✓LEOPARD Syndrome

333. ✓Leonine facies
334. ✓Lizard skin
335. ✓Lobster claw deformity
336. ✓Mauskopf facies
337. ✓Millipede burns
338. ✓Moth-eaten alopecia
339. ✓Moth-eaten borders
340. ✓Moth-eaten appearance of the iris
341. ✓Moth-eaten appearance of skin
342. ✓Moth-eaten scales
343. ✓Monkey face
344. ✓Monkey facies
345. ✓Ostraceous psoriasis
346. ✓Owl's eye appearance
347. ✓Oyster-shell crusts
348. ✓Panda sign
349. ✓Parrot beak nail
350. ✓Parrot beak nose
351. ✓Pigskin appearance
352. ✓Plucked chicken appearance
353. ✓Pseudoparalysis of parrot
354. ✓Rabbit lines'
355. ✓Rabbit burrow appearance
356. ✓Raccoon eyes/owl-eye sign
357. ✓Rat-tail macroconidia
358. ✓Rat-bite fever
359. ✓Rat mite itch
360. ✓Rodent ulcer
361. ✓ Rupioid psoriasis
362. ✓Russell's sign
363. ✓Salmon patch

364. ✓School of fish appearance
365. ✓Sea anemone appearance
366. ✓Shagreen patch
367. ✓Silver–Russell syndrome
368. ✓Snail track ulcers
369. ✓Spider angioma
370. ✓Starfish keratosis
371. ✓Stork bite
372. ✓Swan neck deformity
373. ✓Tadpole cells
374. ✓Tadpole seborrheic keratosis
375. ✓Toad skin
376. ✓Tiger tail pattern
377. ✓Turkey ear
378. ✓Turkey gobbler neck
379. ✓Ugly duckling sign
380. ✓Zebra bodies
381. ✓Zebra-like hyperpigmentation
382. ✓Zebra-stripe pattern
383. Ankyloglossia
384. Ankylostomiasis
385. Annular atrophic plaques of skin (christianson's disease)
386. Annular elastolytic giant cell granuloma AEGCG =Actinic granuloma=Miescher granuloma of the face=O'Brien disease
387. Annular erythema of infancy
388. Annular lichenoid dermatitis of youth
389. Annually recurring erythema annulare centrifugum
390. Anogenital digitate keratosis
391. Anorexia nervosa with Diffuse reticulate purpura DRP

392. **ANOTHER syndrome (alopecia, nail dystrophy, ophthalmic complications, thyroid dysfunction, hypohidrosis, ephelides and enteropathy, and respiratory tract infections)**
393. **Anthrax=Malignant pustule**
394. **Anti-3-hydroxy-3-methylglutaryl coenzyme A reductase immune-mediated necrotizing myopathy**
395. **Antiphospholipid syndrome**
396. **Anti-p105 pemphigoid**
397. **Anti-p200 Pemphigoid= anti-laminin- γ [gamma]1 pemphigoid**
398. **Antithrombin iii deficiency**
399. **Antisynthetase syndrome**
400. **Apert syndrome= acrocephalosyndactyly**
401. **Apical dysplasia of fingers**
402. **Aplasia cutis congenita =Congenital absence of skin : 9 types**
403. **Aphthous ulcer = recurrent aphthous stomatitis (minor & major & herpetiformis)**
404. **Apocrine carcinoma**
405. **Apocrine mixed tumors**
406. **Apocrine nevus**
407. **Aponeurotic fibroma**
408. **Aquagenic pruritus**
409. **Arborizing and stellate melanosis**
410. **Argininosuccinic aciduria**
411. **Argyria**
412. **Arsenicosis**
413. **Arsenic keratosis**
414. **Arsenical melanoderma**
415. **Arterial dissection plus lentiginosis**
416. **Arterial embolism**
417. **Arterial tortuosity syndrome**
418. **Arterio veno lymphatic malformations**
419. **Arteriovenous fistula**
420. **Arteriovenous Haemangioma**

421. **Arteriovenous malformation**
422. **Arthrogryposis**
423. **Arthrogryposis–renal dysfunction–cholestasis (ARC) syndrome**
424. **Ascariasis**
425. **Ascher syndrome**
426. **Aseptic abscess syndrome**
427. **Aspartylglucosaminuria**
428. **Aspergillosis**
429. **Aspirin related diseases**
430. **Astrakhan spotted fever**
431. **Ataxia telangiectasia= louis-bar syndrome**
432. **Athelia**
433. **Athlete's nodule**
434. **Athlete's foot**
435. **Atopic dermatitis /eczema**
436. **Atopic dirty neck**
437. **Atrichia with papular lesion**
438. **Atrophoderma elastolyticum discretum**
439. **Atrophoderma of moulin**
440. **Atrophoderma of pasini and pierini =Sclérodermie atrophique d'emblée=Morphea plana**
441. **Atrophoderma vermiculata**
442. **Follicular atrophoderma**
443. **Atrophia maculosa varioliformis cutis**
444. **Atypical fibroxanthoma= Pseudosarcoma of the skin**
445. **Atypical lichenoid stomatitis**
446. **Atypical lipoma**
447. **Atypical Lipomatous Tumor**
448. **Atypical vascular lesions**
449. **Auricular hematoma**
450. **Australian spotted fever**
451. **Autonomic denervation dermatitis**

452. **Autoimmune/inflammatory syndrome induced by adjuvants (ASIA syndrome)**
453. **Autoimmune lymphoproliferative syndrome (canale–smith syndrome)**
454. **Autoimmune polyendocrinopathy-candidiasis- ectodermal dystrophy) APECED**
455. **Autoimmune polyglandular syndrome : 4Types**
456. **Axillary web syndrome**
457. **Babesiosis**
458. **Bacillary angiomatosis**
459. **Bafverstedt syndrome (benign circumscribed cutaneous lymphadenosis of the areola of the nipple)**
460. **Baisch syndrome**
461. **Baker's cyst**
462. **Balamuthia mandrillaris**
463. **Balanitis**
464. **Balanitis xerotica obliterans**
465. **Balanitis circumscripta plasmacellularis**
466. **Balanoposthitis**
467. **Balloon cell melanoma**
468. **Balloon cell nevus**
469. **Bap-oma**
470. **Bandler syndrome**
471. **Bannayan-riley-ruvalcaba syndrome**
472. **Bannayan-zonana syndrome**
473. **Bannwarth syndrome**
474. **Barber say syndrome**
475. **Bart syndrome (aplasia cutis + epidermolysis bullosa)**
476. **Bartholin duct cyst**
477. **Basaloid follicular hamartoma BFH**
478. **Basal cell carcinoma (see next) BCC**
479. ✓ **Nodular basal cell carcinoma**
480. ✓ **Pigmented type**

481. ✓ **Superficial basal cell carcinoma**
482. ✓ **Morpheaform basal cell carcinoma**
483. ✓ **Fibroepithelial basal cell carcinoma - fibroepithelioma of pinkus**
484. ✓ **Micronodular basal cell carcinoma**
485. ✓ **Cystic basal cell carcinoma**
486. ✓ **Basosquamous carcinoma**
487. ✓ **Large Pore**
488. ✓ **Terebrant carcinoma**
489. ✓ **Polypoid**
490. ✓ **Basal cell nevus syndrome= Nevoid basal cell carcinoma syndrome=Gorlin Goltz syndrome=Gorlin syndrome**
491. ✓ **Multiple hereditary infundibulo-cystic basal cell carcinoma syndrome**
492. **Basan syndrome**
493. **Basomelanocytic tumor**
494. **Bascule syndrome**
495. **Basidiobolomycosis**
496. **Bart-pumphrey syndrome**
497. **Bateman purpura**
498. **Bateman dermatitis**
499. **Battered child syndrome**
500. **Battered baby syndrome**
501. **Bazex- dubre- christol syndrome**
502. **Bazex syndrome**
503. **BCOR-CCNB3 undifferentiated sarcomas**
504. **Beals–Hecht syndrome= contractural arachnodactyly**
505. **Bean syndrome**
506. **Beare–stevenson syndrome**
507. **Betel leaf (piper betle)**
508. **Beckwith–wiedemann syndrome=Exomphalos-macroglossia -gigantism syndrome**
509. **Behcet disease**
510. **Bejel related disases=Endemic Syphilis**

511. **Bendar tumour**
512. **Benign alveolar ridge keratosis**
513. **Benign Follicular Hamartoma**
514. **Benign lymphoepithelial cyst**
515. **Benign mucinous metaplasia of the genitalia**
516. **Benign neonatal hemangiomatosis**
517. **Benign symmetric lipomatosis**
518. **Benign triton tumor =neuromuscular hamartoma =choristoma**
519. **Berardinelli's syndrome**
520. **Berlin syndrome**
521. **Berloque dermatitis= photo toxic contact dermatitis**
522. **Berloque dermatitis(freckle like)**
523. **Berk-tabatznik syndrome**
524. **Besnier prurigo**
525. **Bicyclist's vulva**
526. **Bier spot =Angiospastic macules**
527. **Bilharziasis = Schistosomiasis**
528. **Biotin deficiency**
529. **Biotinidase deficiency**
530. **Bird nest hair**
531. **Bird-headed dwarfism= seckel syndrome**
532. **Birt-hogg-dubé syndrome(alias Hornstein-Knickenberg syndrome)**
533. **Bites&stings&venoms(see next)**
534. **✓Arthropod bite**
535. **✓Anemon**
536. **✓Bee**
537. **✓Rove beetle**
538. **✓Blister beetles/Rove beetle**
539. **✓lepidopterism**
540. **✓Bug**
541. **✓Cat**
542. **✓Caterpillar**

543. ✓Centipedes
544. ✓Chigger
545. ✓Ciguatera
546. ✓ Coral
547. ✓Dog
548. ✓Dogger Bank itch
549. ✓Fire ant
550. ✓Fish
551. ✓Flea
552. ✓Jelly fish
553. ✓Leeches
554. ✓Lizard
555. ✓Locusta
556. ✓marine injuries
557. ✓Mite
558. ✓Mollusca (Octopuses and Conus Snails)
559. ✓Mosquito
560. ✓Portguese man of war =physalia physalis
561. ✓Sea bather ’s eruption
562. ✓Scorpion
563. ✓Snake
564. ✓ Spider
565. ✓Loxosceles Spiders
566. ✓Widow Spiders
567. ✓Tarantulas
568. ✓Tick
569. ✓Tongueworm
570. ✓Urchin
571. ✓Wasps
572. ✓Weever
573. Bizarre Parosteal Osteochondromatous Proliferation

574. **Björnstad syndrome**
575. **Adult Blaschkitis**
576. **Acquired blaschkoid dermatitis**
577. **Blaschko linear acquired inflammatory skin eruption (BLAISE)**
578. **Blastomycosis=North american blastomycosis=Gilchrist's Disease**
579. **Blau syndrome=juvenile systemic granulomatosis**
580. **Blepharochalsis**
581. **Blepharophyma**
582. **Blepharo-cheilo-dontic syndrome**
583. **Blistering dactylitis**
584. **Bloch-sulzberger syndrome**
585. **Bloom syndrome=congenital telangiectatic erythema and stunted growth**
586. **Blue baby syndrome**
587. **Blueberry muffin baby**
588. **Blue man syndrome**
589. **Blue nevus**
590. **Blue nevus-like metastatic melanoma**
591. **Blue rubber bleb nevus syndrome= bean syndrome**
592. **Blue toe syndrome**
593. **Blue tongue**
594. **Bockenheimer syndrome**
595. **Body dysmorphic disorder**
596. **Bohn nodule**
597. **Boil**
598. **Bonnet–Dechaume–Blanc syndrome**
599. **Book dysplasia**
600. **Book syndrome=Hyperhidrosis, premature canities and premolar aplasia**
601. **Börjeson–forssman–lehmann syndrome**
602. **Borrelial lymphocytoma cutis**
603. **Borrone dermato-cardio-skeletal syndrome**
604. **Botryomycosis=Bacterial pseudomycosis**
605. **Bowel-associated dermatosis–arthritis syndrome BADAS**
606. **Bourneville-pringle syndrome**

607. **Boutonneuse fever =Mediterranean Spotted Fever**
608. **Bowen disease=squamous cell carcinoma in situ**
609. **Bowenoid papulosis**
610. **Bowenoid actinic keratosis**
611. **Bpes (blepharophimosis, ptosis, and epicanthus inversus syndrome (BPES)**
612. **Brachioradial pruritus**
613. **Brachydactyly with absence of middle phalanges and hypoplastic nails or brachydactyly type A5 with nail dysplasia**
614. **Brachymetapody–anodontia–hypotrichosis–albinoidism(oculo-osteocutaneoussyndrome; tuomaala syndrome**
615. **Brachymorphism-onychodysplasia-dysphalangism(BOD)**
616. **Branched-chain amino acid deficiency**
617. **Branchial cleft ,sinuses, and fistulae**
618. **Branchial Cleft cyst = Lymphoepithelial cyst= Lateral cervical cyst**
619. **Branchio-oculo-facial syndrome**
620. **Branchio-oto-renal syndrome**
621. **Brazilian hemorrhagic fever**
622. **BRESEK/BRESHECK syndrome(Brain anomalies-retardation of mentality and growth-ectodermal dysplasia-skeletal malformations-Hirschprung disease-ear deformity and deafness-eye hypoplasia-cleft palate-cryptorchidism-kidney dysplasia/hypoplasiaBromhidrosis**
623. **Bromidrosiphobia**
624. **Bromoderma**
625. **Bronchogenic cyst**
626. **Bronze baby syndrome**
627. **Brooke-spiegler syndrome**
628. **Brucellosis=Undulant fever**
629. **Bruton syndrome**
630. **Bubble hair**
631. **Buccal exostoses**
632. **Buerger diseases**
633. **Buffalopox**
634. **Bulimia**

635. **postgraft Blisters**
636. **Bulla after burn**
637. **Coma bulla=Neurologic blisters**
638. **Bullous disease of diabetes=Bullosis diabeticorum**
639. **Edema Bulla= Hydrostatic bullae**
640. **Fracture bulla**
641. **Friction blister**
642. **Bullous hemorrhagic dermatosis**
643. **Bullous dermatitis artefacta**
644. **Bullous drug reactions**
645. **Bullous insect bite**
646. **Bullous neutrophilic dermatosis**
647. **Bullous urticarial**
648. **Bullous pemphigoid (see next)**
649. ✓ **Juvenile bullous pemphigoid**
650. ✓ **Brunsting–perry form**
651. ✓ **Dyshidrosiform pemphigoid**
652. ✓ **Eczematous pemphigoid**
653. ✓ **Erythrodermic pemphigoid**
654. ✓ **Large erosive ten-like lesions**
655. ✓ **Lichen planus pemphigoides**
656. ✓ **localized lesions**
657. ✓ **Papular pemphigoid**
658. ✓ **Pemphigoid nodularis**
659. ✓ **Pemphigoid vegetans**
660. **Bureau–barrière syndrome**
661. **Burn**
662. **Cigarette burns**
663. **Burn Hemangioma (BH) =Scalded Pyogenic Granuloma**
664. **Aspirin alcohol,acrylic resin,iodine : burn of oral cavity**
665. **Burning feet syndrome**
666. **Burning mouth syndrome**

667. **Burning scalp syndrome**
668. **Burning scrotum syndrome**
669. **Buschke-lowenstein tumour**
670. **Buschke ollendorff syndrome**
671. **Bushy eyebrows**
672. **Byars-jurkiewicz syndrome**
673. **Cactus granuloma**
674. **Cadasil syndrome (cerebral autosomal dominant Arteriopathy with subcortical infarcts and leuko-encephalopathy)**
675. **CAHMR syndrome (cataract, hypertrichosis, mental retardation)**
676. **Café-au-lait macules CALM**
677. **Cafe noir**
678. **Calabar swelling**
679. **Calcinosis circumscripta**
680. **Calcinosis universalis**
681. **Tumoral calcinosis**
682. **Calcinosis cutis (metastatic & dystrophic & idiopathic & iatrogenic & calcinosis universalis)**
683. **Calculus**
684. **Calciphylaxis= calcific uremicarteriolopathy=Calcifying Panniculitis**
685. **Calcifying aponeurotic fibroma**
686. **Calcifying fibrous pseudo-tumour**
687. **Caliber-persistent artery**
688. **Callosities**
689. **Callus**
690. **Camarena syndrome**
691. **Camelpox**
692. **Canalicular adenoma**
693. **Cancrum oris= noma= Necrotising stomatitis**
694. **Candida = candidiasis = candidosis**
695. ✓ **Angular cheilitis (perlèche)**
696. ✓ **Thrush (acute pseudo-membranous candidiasis)**

697. ✓ Chronic pseudomembranous candidiasis
698. ✓ Candidal leucoplakia (chronic hyperplastic candidiasis)
699. ✓Acute atrophic candidosis= Acute antibiotic stomatitis
700. ✓ Acute erythematous candidosis
701. ✓ Chronic denture stomatitis (chronic atrophic candidiasis)
702. ✓Chronic erythematous candidosis
703. ✓ Chronic mucocutaneous candidiasis
704. ✓Chronic nodular candidosis
705. ✓Candida napkin/nappy dermatitis
706. ✓Antibiotic (iatrogenic) candidiasis
707. ✓Cheilocandidiasis
708. ✓Congenital cutaneous candidiasis
709. ✓Neonatal candidiasis
710. ✓Candidal vulvovaginitis
711. ✓Candidal Balanitis
712. ✓Candidal intertrigo=Flexural Candidiasis
713. ✓Candidal Miliaria
714. ✓Candidal Granuloma
715. ✓Candidid
716. ✓Candidemia
717. ✓Acute candidosis(Thrush& Acute antibiotic stomatitis)
718. ✓Chronic candidosis(Denture-induced stomatitis&Chronic hyperplastic candidosis&Chronic mucocutaneous candidosis&Erythematous candidosis)
719. CANDLE: chronic atypical neutrophilic dermatosis with lipodystrophy and elevated temperature
720. Canities (premature canities &canities subita)
721. Cannon disease= white sponge naevus
722. Cantu syndrome= hypertrichotic osteochondrodysplasia
723. Capillary leak syndrome
724. Capillary malformation – arteriovenous malformation syndrome(CM-AVM syndrome)

725. **Capnocytophaga canimorsus bacteremia**
726. **Carboxylase deficiency**
727. **Carbuncle**
728. **Carcinoid syndrome**
729. **Carcinosarcoma=Metaplastic carcinoma of the skin =Sarcomatoid Carcinoma**
730. **Carcinoma en cuirasse**
731. **Carcinoma erysipeloides**
732. **Carcinoma telangiectoides**
733. **Card14-mediated pustular psoriasis CAMPS**
734. **CARD-14 associated papulosquamous eruption (CAPE)**
735. **Cardio-acro-facial syndrome= rabenhorst's syndrome**
736. **Cardiofaciocutaneous syndrome**
737. **Cardiomyopathy with alopecia and palmoplantar keratoderma (CAPK)**
738. **Carey syndrome**
739. **Carney complex**
740. **Carnitine translocase deficiency**
741. **Carotenemia**
742. **Carpenter syndrome**
743. **Cartilage hair hypoplasia= mukusick-type metaphyseal chondrodysplasia**
744. **Cartilaginous papule of ear**
745. **Cartilagnous rest of neck**
746. **Carrion disease**
747. **Castleman diseases (giant lymph node hyperplasia, angiofollicular lymph node hyperplasia)**
748. **Cat scratch disease**
749. **Cataract–alopecia–sclerodactyly syndrome**
750. **Cattlepox**
751. **Cavernous hemangioma**
752. **Cavernous lymphangioma**
753. **CEDNIK: cerebral dysgenesis, neuropathy, ichthyosis, and keratoderma**
754. **Cellulite= gynoid lipodystrophy**
755. **Cellulitis**

756. **Helicobacter cellulitis**
757. **Cerebello -Trigeminal - Dermal dysplasia**
758. **Cerebro-Dermato-Osseous-Dysplasia**
759. **Cerebro-oculo-facio-skeletal (COFS)**
760. **Ceruminous adenoma**
761. **Ceruminous carcinoma**
762. **Cervical chondrocutaneous branchial remnants**
763. **Cysticercosis:Cestodes infections (Taenia saginata&Taenia solium)**
764. **Chagas disease= American trypanosomiasis**
765. **Chancre of syphilis**
766. **Chancroid=soft chancres**
767. **CHANDS (curly hair, ankyloblepharon,nail dysplasia)syndrome**
768. **Charcot-marie-tooth**
769. **CHARGE syndrome = (coloboma, heart defects, atresia choanae, Growth retardation, genital abnormalities, and ear abnormalities)**
770. **Chediak - higashi syndrome (CHS) =Béguez César–Steinbrinck syndrome**
771. **Angular cheilitis=angular stomatitis=perleche**
772. **Actinic cheilitis=Solar Cheilosis**
773. **Atopic cheilitis**
774. **Contact cheilitis**
775. **Drug induced cheilitis**
776. **Exfoliative cheilitis**
777. **Factitial cheilitis**
778. **Granulomatous cheilitis=miescher's cheilitis**
779. **Glandular cheilitis (see next)**
780. **✓Cheilitis Glandularis Apostematosa=Volkmann's cheilitis**
781. **✓superficial suppurative = Baelz-Unna**
782. **✓Glandular cheilitis simplex**
783. **Infective cheilitis**
784. **Nutritional cheilitis**
785. **Plasma cell cheilitis**
786. **Cheilitis simplex**

787. **Lip Chapping**
788. **Cheiralgia paresthetica**
789. **Cheiroarthropathy**
790. **Cherry angioma=cherry hemangioma=senile angioma=campbell–demorgan spot**
791. **Cherubism**
792. **Cheyletiella dermatitis**
793. **Chickenpox =varicella**
794. **Chickenpox scar**
795. **Neonatal varicella**
796. **Varicella gangrenosum**
797. **Chickenpox: photolocalized type**
798. **Chickenpox bullosa**
799. **Chikungunya- chicku**
800. **Childhood flexural comedones**
801. **Chlorotrichosis=green hair**
802. **Chondrodermatitis nodularis helics**
803. **Chondrodermatitis nodularis nasi (of nose)**
804. **Chondromas= Soft Tissue Chondroma**
805. **Chondroblastic osteosarcoma**
806. **Chloroma=Granulocytic sarcoma**
807. **Chondroid lipoma**
808. **Chondroid syringoma= mixed tumor of the skin**
809. **Malignant chondroid syringoma**
810. **Chondrosarcoma**
811. **chromhidrosis**
812. **Chromobacteriosis**
813. **Chromoblastomycosis=Chromomycosis=Fonseca's disease**
814. **Chronic bullous disease of childhood CBDC = linear IGA disease of childhood**
815. **Chronic eczematous eruptions of the aged**
816. **Chronic granulomatous disease**
817. **Chronic Idiopathic Penile edema**

818. **Chronic infantile neurological cutaneous and articular syndrome CINCa**
819. **Chronic recurrent annular neutrophilic dermatosis**
820. **Chronic nipple eczema**
821. **Chronic non-bacterial osteomyelitis**
822. **Chronic recurrent multifocal osteomyelitis**
823. **Chronic ulcerative stomatitis**
824. **Chrysiasis**
825. **Churg strausse vasculitis= Eosinophilic granulomatosis with polyangiitis=Allergic angiitis and granulomatosis**
826. **Cicatricial pemphigoid= Mucous Membrane Pemphigoid**
827. **Ciliated cyst of the vulva**
828. **Circle hairs**
829. **Circumscribed Palmar or plantar hypokeratosis**
830. **Circumferential skin creases Kunze type**
831. **Cisriod aneurysm**
832. **Citrullinemia**
833. **CLAPO (capillary malformation of the lower lip, lymphatic malformation predominant on the face and neck, asymmetry, and partial=generalized overgrowth)**
834. **Clarkson's syndrome**
835. **Clear cell acanthoma - acanthoma of degos**
836. **Clear cell hidradenocarcinoma**
837. **Clear cell papulosis**
838. **Clear cell squamous cell carcinoma**
839. **Clear cell syringoma**
840. **Cleft lip**
841. **Cleft palate**
842. **Cleft palate–lateral synechia syndrome**
843. **Cleidocranial dysostosis with micrognathia, absent thumbs**
844. **Cleidocranial dysostosis with micrognathia, absent thumbs, and distal aphangia, Yunis–Varon syndrome**
845. **Cloacogenic carcinoma**

846. **CLOVES syndrome (Congenital, Lipomatous, Overgrowth, Vascular Malformations, Epidermal Nevi and Spinal/Skeletal Anomalies and/or Scoliosis))**
847. **Coagulopathies**
848. **Cobb syndrome=cutaneomeningospinal angiomatosis**
849. **Coccidioidomycosis= San Joaquin valley fever= Posadas' Disease**
850. **Coccydynia**
851. **Cockayne syndrome**
852. **Coenurosis**
853. **Coffin–siris syndrome**
854. **Coffin-Lowry syndrome**
855. **Cogan syndrome**
856. **Cohen syndrome**
857. **Coin-rubbing injuries**
858. **Cold agglutinin disease**
859. **Cold injury**
860. **Cole syndrome**
861. **Collagenous fibroma**
862. **Eruptive Collagenoma**
863. **Familial cutaneous Collagenoma**
864. **Isolated/solitary collagenoma**
865. **Collagenosis nuchae**
866. **Collagenous Papule of the Eyelid**
867. **Plantar cerebriform collagenoma**
868. **Papulolinear collagenoma=linear collagen nevus**
869. **Collagenous and elastotic plaques of the hands=degenerative collagenous plaques of the hands= keratoelastoidosis marginalis**
870. **Colloid milium= Elastosis colloidalis conglomerata=Miliary colloidoma**
871. **Collision tumour**
872. **Colorado tick fever**
873. **Comedocarcinoma**
874. **Comedo-like acantholytic dyskeratosis of the face and scalp**
875. **Open comedones**

876. **Microcomedone**
877. **Giant comedone**
878. **Macrocomedone**
879. **Submarine comedones**
880. **Sandpaper comedones**
881. **Double-edged comedones**
882. **Polyporous comedones**
883. **Compartment syndrome**
884. **Complement deficiency**
885. **Complex regional pain syndrome= reflex sympathetic dystrophy**
886. **Composite hemangioendotheliomna**
887. **Compulsuive hand washing**
888. **Coenurosis**
889. **Condyloma acuminatum**
890. **Condyloma lata**
891. **Confetti-like macular atrophy**
892. **Confluent and reticulated papillomatosis gougerot-carteaud**
893. **Congenital cd34+ granular cell dendrocytosis**
894. **Congenital comedones**
895. **Congenital deficiency of glutamine synthetase**
896. **Congenital diffuse melanosis**
897. **Congenital disorders of glycosylation**
898. **Congenital disseminated comedones**
899. **Congenital epulis / Epulides**
900. **Congenital erosive and vesicular dermatosis**
901. **Congenital fibrous hamartoma**
902. **Congenital hereditary endothelial dystrophy with nail hypoplasia**
903. **Congenital hypertrichotic melanoneurocytoma (CHM)**
904. **Congenital idiopathic generalized anhidrosis (CIGA)**
905. **Congenital infiltrating lipomatosis**
906. **Congenital ingrown toenails**
907. **Congenital insensitivity to pain =Hereditary sensory and autonomic neuropathies (HSAN)**

908. **Congenital linear atrophoderma**
909. **Congenital localized scleroderma**
910. **Congenital midline Cervical cleft**
911. **Congenital Perineal Groove**
912. **Congenital peristernal dermal sinuses**
913. **Congenital pseudarthrosis of the clavicle**
914. **Congenital rubella syndrome**
915. **Congenital subungual pterygium**
916. **Congenital trichoid keratosis**
917. **Congenital vaccinia**
918. **Congenital varicella syndrome (CVS)**
919. **Congenital venous malformation**
920. **Conidobolus coronatus related Infections**
921. **Contact stomatitis**
922. **Cooks syndrome**
923. **Copper (toxicity&deficiency)**
924. **COPS (calcinosis cutis, osteoma cutis, poikiloderma, and skeletal abnormalities)**
925. **Corn**
926. **Cornelia de lange syndrome=typus amstelodamensis =amsterdam dwarf=Brachmann-de Lange Syndrome**
927. **Corneodermato -osseous syndrome**
928. **Corona alopecia**
929. **Corona seborrhoeica**
930. **Corona veneris**
931. **Corona psoriatica**
932. **Costal fringe**
933. **Costello syndrome =faciocutaneoskeletal syndrome**
934. **Covid 19 (toes&fingers&COVID arm&COVID nails&COVID "red half‐moon nail" sign &COVID tongue&COVID mask&Kawa‐COVID‐19&COVID rash& COVID hand dermatitis)**
935. **Cowden syndrome**
936. **Cowpox**

937. Coumadinn (coumarin, warfarin)-induced necrosis
938. Cranio ectodermal dysplasia=Sensenbrenner Syndrome
939. Craniofacial dysostosis with hypertrichosis and hypodontia
940. Craniofrontonasal dysplasia
941. Craniofaciocutaneous syndrome
942. Craniomandibular Dermatodysostosis =Familial Mandibulo-Acral Dysplasia
943. Craniosynostosis
944. Craniosynostosis ,anal anomalies and porokeratosis CAP syndrome
945. CREST syndrome (calcinosis, Raynaud phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia)
946. Cri-du-chat syndrome
947. Crimean congo hemorrhagic fever
948. Crohn disease
949. Metastatic Crohn's disease
950. Cronkhite–Canada syndrome
951. Cross syndrome
952. Crouzon syndrome = craniofacial dysostosis
953. Crow's feet
954. Crow-fukase syndrome
955. CRTC1::TRIM11 cutaneous tumours (CTCT)
956. Cryoglobulinemia (3 types)
957. Cryofibrinogenemia
958. Crystalglobulin vasculopathy
959. Cryopyrin-associated periodic syndromes (CAPS)
960. Cryotherapy related complications
961. Cryptococcosis= European Blastomycosis
962. Cupping injuries
963. cutaneous arteriolitis
964. Cutaneous collagenous vasculopathy
965. Cutaneous ciliated cyst of lower limbs
966. Cutaneous endometriosis

Cutaneous eruption associated with streptococcus dysgalactiae bacteremia

967. **Cutaneous granulocytic sarcoma**
968. **Cutaneous heterotopic bronchogenic tissue**
969. **Cutaneous horn=cornu cutaneum**
970. **Cutaneous lymphadenoma**
971. **Cutaneous metallosis**
972. **Cutaneous microsphaeropsis arundinis**
973. **Cutaneous neosartorya hiratsukae infection**
974. **Cutaneous paecilomyces infection**
975. **Cutaneous T-cell dyscrasia (CTCD)**
976. **Cutaneous waldenström macroglobulinemia**
977. **Cutaneous yarrowia lipolytica infection**
978. **Cutis laxa=dermatochalasia= dermatomegaly =generalized elastolysis =generalized elastorrhexis(see next)**
979. **✓ADCL**
980. **✓Progeroid ADCL (PADCL)**
981. **✓ARCL type 1A**
982. **✓ARCL type 1B**
983. **✓ARCL type 1C(Urban–Rifkin–Davis syndrome)**
984. **✓ARCL type 2A (Debré type; allelic to wrinkly skin syndrome)**
985. **✓ARCL type 2B**
986. **✓ARCL types 2C and 2D**
987. **✓De Barsy syndrome A (ARCL type 3A)**
988. **✓De Barsy syndrome B (ARCL type 3B)**
989. **Cutis marmorata**
990. **Cutis marmorata telangiectatica congenital=van Lohuizen syndrome**
991. **Cutis pleonasmus**
992. **Cutis rhomboidalis nuchae**
993. **Cutis tricolor**
994. **Cutis verticis gyrate= pachydermia verticis gyrate**
995. **Curry-jones syndrome**
996. **Cyanosis**
997. **Cyclic edema**

998. **Cyclical inguinal keratoderma**
999. **Cydnidae =burrowing bug**
1000. **Cylindroma**
1001. **Cylindrocarcinoma=malignant cylindroma**
1002. **Cyrano nose**
1003. **Cystadenoma**
1004. **Cystathionine synthetase deficiency**
1005. **Cysticercosis**
1006. **Cystic Condyloma**
1007. **Cystic fibrosis**
1008. **Cystic Neutrophilic Granulomatous Mastitis (CNGM)**
1009. **Cytomegalovirus infection (CMV)**
1010. **D-2-hydroxyglutaric aciduria**
1011. **Dabska's tumour**
1012. **Dacrocystitis**
1013. **Dadru**
1014. **Dandruff= pityriasis simplex capillitii**
1015. **Danthron erythema**
1016. **Darier white = dyskeratosis follicularis =psorospermose folliculaire vegetante (see next)**
1017. **✓Guttate hypopigmentation**
1018. **✓Hyperpigmented flat-topped papules and macules=freckle-like**
1019. **✓Vesiculobullous lesions**
1020. **✓Acral hemorrhagic lesions**
1021. **✓Keratoderma**
1022. **✓ Cornifying**
1023. **✓Comedones**
1024. **✓Periocular nodulocystic lesions**
1025. **✓Alopecia**
1026. **✓cutis verticis gyrata**
1027. **Darier–roussy disease**
1028. **Pseudo-darier's sign**

1029. **Dark halo/circles**
1030. **Davener's dermatosis (DD)**
1031. **Deafness and ear pits**
1032. **Deafness, with vitiligo and muscle wasting**
1033. **De barsy syndrome**
1034. **Deep benign fibrous histiocytoma**
1035. **Deep venous thrombosis**
1036. **Defects of the interleukin-12/interferon-γ axis**
1037. **Defects in Toll-like receptor (TLR) signaling**
1038. **Deficiency of adenosine deaminase 2 (DADA2)**
1039. **Deficiency of interleukin-1 receptor antagonist DIRA**
1040. **Deficiency of interleukin-36 receptor antagonist DITRA**
1041. **Defoliants**
1042. **Degos disease = malignant atrophic papulosis**
1043. **Delleman syndrome**
1044. **Delusion parasitosis=ekbom syndrome= Delusional infestation**
1045. **Delusional hyperhidrosis**
1046. **Demodicosis &demodex (see next)**
1047. **✓Primary**
1048. **✓Secondary**
1049. **✓ pityriasis folliculorum=Facial Spinulate Demodicosis**
1050. **✓ Periorificial dermatitis Demodicosis**
1051. **✓ Facial dermatitis**
1052. **✓Demodex folliculitis**
1053. **✓Nodulocystic/conglobate demodicosis**
1054. **✓ papulopustular demodicosis**
1055. **✓ Demodex abscesses**
1056. **✓ Ocular Demodicosis**
1057. **✓Auricular Demodicosis**
1058. **✓Pigmented demodicosis**
1059. **Dengue fever**
1060. **Dento-oculocutaneous syndrome(ackerman syndrome)**

1061. **Denture associated fibrous hyperplasia**
1062. **Denture stomatitis**
1063. **De sanctis–cacchione syndrome of xeroderma pigmentosa**
1064. **Dental caries**
1065. **Dental sinus =sinus of dental origin**
1066. **Dependency disease**
1067. **Dependent edema**
1068. **Dercum disease =Adiposis Dolorosa**
1069. **Dermal dendrocyte hamartomas**
1070. **Dermal dendrocyte hamartoma with stubby white hair**
1071. **Dermal duct tumour**
1072. **Dermal hyperneury**
1073. **Dermal melanocytic Hamartoma**
1074. **Dermatitis factitial = artifacta**
1075. **Dermatitis cruris pustulosa atrophican=folliculitis decalvans cruris=Sycosis Cruris**
1076. **Dermatitis herpetiformis DH= Duhring 's disease**
1077. **Dermatitis medicamentosa**
1078. **Dermatitis neglecta/Dermatosis neglecta (DN)**
1079. **Dermatitis palmaris sicca**
1080. **Dermatitis passivata**
1081. **Dermatitis plantaris sicca**
1082. **Dermatitis repens**
1083. **Dermatitis simulata**
1084. **Dermatitis vegetans**
1085. **Dermatitis venenata**
1086. **Dermatodaxia**
1087. **Dermatofibroma =benign fibrous histiocytoma= fibroma simplex= sclerosing haemangioma (see next)**
1088. **✓Aneurysmal dermatofibroma**
1089. **✓Ankle-type dermatofibroma (lipidized fibrous histiocytoma)**
1090. **✓Annular hemosiderotic dermatofibroma**

1091. ✓Delled (“atrophic”) dermatofibroma
1092. ✓Dermatofibroma with satellitosis
1093. ✓Disseminated multiple dermatofibromas
1094. ✓Erosive and ulcerated dermatofibroma
1095. ✓Giant dermatofibroma
1096. ✓Hyperkeratotic dermatofibroma
1097. ✓Hyperpigmented nodular dermatofibroma
1098. ✓Multinodular hemosiderotic dermatofibroma
1099. ✓Multiple grouped dermatofibromas
1100. ✓Nonpigmented dermatofibroma
1101. ✓Pedunculated or polypoid dermatofibroma
1102. ✓Subcutaneous dermatofibroma
1103. Dermatofibrosis lenticularis disseminate
1104. Dermatofibrosarcoma protuberans
1105. Pigmented dermatofibrosarcoma protuberans (bednar)
1106. Congenital abnormalities of dermatoglyphs(ridge aplasia, ridge hypoplasia, ridge dissociation, ridges-off-the-end)
1107. Dermatographism **(see next)**=Dermatographia=Factitious Urticaria
1108. ✓ Black dermatographism
1109. ✓ Cholinergic dermatographism
1110. ✓ Red dermatographism
1111. ✓ White dermatographism
1112. ✓ Yellow dermatographism
1113. ✓Cold precipitated dermographism
1114. ✓Delayed dermographism
1115. ✓Follicular dermographism
1116. ✓Immediate dermographism
1117. Dermatoheliosis
1118. Dermatomycosis
1119. Dermatomyofibroma

1120. **Dermatomyositis DM (see next)**
1121. **✓ Adult-onset**
1122. **✓ Juvenile onset**
1123. **✓Classic**
1124. **✓ Amyopathic**
1125. **✓Hypomyopathic**
1126. **✓Clinically amyopathic**
1127. **✓Classic dm with malignancy**
1128. **✓Dermatomyositis sine myositis**
1129. **Dermatophytids**
1130. **Dermatopathia pigmentosa reticularis (DPR)**
1131. **Dermatophagia**
1132. **Dermatoporosis**
1133. **Dermatosis papulosis nigra DPN**
1134. **Dermatorynchus geneae**
1135. **Dermestid dermatitis =carpet beetle dermatitis**
1136. **Dermacentor-borne necrosis erythema and lymphadenopathy (DEBONEL)**
1137. **Dermochondrocorneal syndrome= françois'syndrome**
1138. **Dermo-odontodysplasia**
1139. **Dermoid cyst**
1140. **Desmons syndrome**
1141. **Diabetic/ diabetes related disorders**
1142. **Diabetic bullae (Bullous diabeticorum)**
1143. **Diabetic dermopathy**
1144. **Diabetic Thick Skin =Cheiroarthropathy**
1145. **Didymosis aplasticosebacea**
1146. **Diffuse capillary malformation with overgrowth DCMO**
1147. **Diffuse dermal angiomatosis**
1148. **Diffuse lipomatosis**
1149. **Digeorge's syndrome**
1150. **Digital Fibromatosis**
1151. **Digital papillary adenocarcinoma DPA**

1152. **Digital papular calcinosis**
1153. **Digital papular calcific elastosis**
1154. **Digital verrucous fibroangioma**
1155. **Digitalgia paresthetica**
1156. **Dilated pore of winer**
1157. **Dimple & skin fossae**
1158. **Diogenes syndrome**
1159. **Dipetalonemiasis**
1160. **Diphtheria**
1161. **Dirofilariasis**
1162. **Dissecting cellulitis= folliculitis capitis abscedens et suffodiens**
1163. **Disseminated hypopigmented keratoses (DHK)**
1164. **Disseminated intravascular coagulation (DIC)**
1165. **Distichiasis**
1166. **Divry-van bogaert syndrome= cortico-meningeal angiomatosis**
1167. **Dock 8 deficiency :dedicator of cytokinesis 8**
1168. **Donohue syndrome**
1169. **Donovanosis= granuloma inguinale**
1170. **Doors syndrome(deafness, onychodystrophy, osteodystrophy, and mental retardation)**
1171. **Dowling degos disease**
1172. **Down syndrome**
1173. **Vesiculopustular eruption of transient myeloproliferative disorder in down syndrome**
1174. **Double eyebrow**
1175. **Dracunculiasis=guinea worm disease= Dracontiasis**
1176. **Drug abuse**
1177. **Cutaneous adverse drug reactions**
1178. **Bullous drug reactions**
1179. **Drug exanthema - morbilliform drug eruption= maculopapular drug eruptions**
1180. **Drug reaction with eosinophilia and systemic symptoms DRESS**
1181. **Drug-associated bullous pemphigoid**

1182. **Drug-associated linear iga dermatosis**
1183. **Drug-induced Acanthosis nigricans**
1184. **Drug-induced Accelerated rheumatoid nodulosis**
1185. **Drug-induced Acne (DIA)**
1186. **Drug-induced Acneiform eruption (DIAE)**
1187. **Drug-induced Alopecia**
1188. **Drug-induced Erythroderma**
1189. **Drug-Induced Linear Iga Disease**
1190. **Drug-induced lupus erythematosus**
1191. **Drug-induced hyperpigmentation**
1192. **Drug-induced hypopigmentation**
1193. **Drug induced changes in hair color**
1194. **Drug-induced hirsutism**
1195. **Drug-induced hypertrichosis**
1196. **Drug-induced ichthyosis**
1197. **Drug-induced interstitial granulomatous**
1198. **Drug-induced nail abnormalities**
1199. **Drug-induced necrotizing purpura**
1200. **Drug-induced Palmoplantar keratoderma**
1201. **Drug-induced pemphigus**
1202. **Drug triggered pemphigus**
1203. **Drug-induced Photoallergic reaction**
1204. **Drug-induced phototoxic reaction**
1205. **Drug-induced pruritus**
1206. **Drug-induced pseudolymphoma= lymphomatoid drug reaction**
1207. **Drug-Induced Pseudoporphyria**
1208. **Drug-induced sarcoidosis**
1209. **Drug induced scarring hair loss**
1210. **Drug-induced sweet's syndrome**
1211. **Drug-induced vasculitis**
1212. **Drug-induced vitiligo**
1213. **Anaphylactoid reaction= Kounis syndrome**
1214. **Fixed drug eruption FDE**

1215. **Injection site reaction**
1216. **Lichen planus pigmentosus (lpp) like eruption**
1217. **Lichenoid drug eruption**
1218. **Palifermin-associated papular eruption**
1219. **Pityriasiform drug eruption**
1220. **psoriasiform drug eruption**
1221. **Purpuric drug reactions**
1222. **Pustular drug reactions**
1223. **Scleroderma-like reactions to taxanes**
1224. **Serum sickness-like reaction**
1225. **Symmetrical drug-related intertriginous and flexural exanthema = Baboon syndrome**
1226. **True serum sickness**
1227. **Chemotherapy related cutanous manifestation**
1228. **✓Acral erythrodysesthesia**
1229. **✓Ara -c ear**
1230. **✓Burgdorf's reaction**
1231. **✓Chemotherapy- -induced oral mucositis**
1232. **✓Chemotherapy recall dermatitis**
1233. **✓Eccrine squamous syringometaplasia**
1234. **✓Epidermal dysmaturation**
1235. **✓Epidermal dystrophy**
1236. **✓Erythrodysesthesia**
1237. **✓Extravasation**
1238. **✓Flexural erythematous eruption**
1239. **✓Hand foot skin reaction**
1240. **✓Hand foot skin syndrome**
1241. **✓Intertriginous eruption associated with chemotherapy**
1242. **✓Intertrigo-like eruption, chemotherapy induced**
1243. **✓Palmar-plantar erythema**
1244. **✓Palmar-plantar erythrodysesthesia**
1245. **✓Toxic acral erythema**

1246. ✓Toxic erythema of chemotherapy
1247. ✓Toxic erythema of the palms and soles

Drugs related cutanous manifestation:

1248. ✓Allopurinol hypersensitivity syndrome
1249. ✓Arsenic
1250. ✓BCR-ABL Tyrosine Kinase Inhibitors
1251. ✓Bleomycin
1252. ✓BRAF inhibitor
1253. ✓Clofazimine
1254. ✓Dapsone hypersensitivity syndrome
1255. ✓Epidermal growth factor inhibitors
1256. ✓Hydroquin
1257. ✓Insulin
1258. ✓Mitogen-activated protein kinase (MAPK) inhibitors
1259. ✓MEK1 inhibitor
1260. ✓Mercury induced skin lesions
1261. ✓Methotrexate
1262. ✓Methotrexate (MTX) toxicity
1263. ✓Minocycline hypersensitivity syndrome
1264. ✓Minoxidil
1265. ✓Proteasome inhibitors
1266. ✓Retinoid
1267. ✓Steroid
1268. Dubowitz syndrome
1269. Dupuytren contracture = Palmar Fibromatosis
1270. Dwarfism–alopecia–pseudoanodontia–cutis laxa
1271. Dwarfism, cerebral atrophy and keratosis pilaris
1272. Acquired Brachial Cutaneous Dyschromatosis
1273. Dyschromatosis symmetrica hereditaria
1274. Dyschromatosis universalis hereditaria
1275. Dyssebacea

1276. **Dyskeratosis congenita (zinsser–engman–cole syndrome)**
1277. **Dysthesia**
1278. **Dysesthetic anogenital pain syndromes**
1279. **Ear pit =preauricular sinus**
1280. **Easy bruising syndrome=Inherited giant platelet disorders(Bernard–Soulier syndrome&Velocardial syndrome & Montreal platelet syndrome&May–Hegglin anomaly &Gray platelet syndrome&Sebastian syndrome&Epstein syndrome&Fechtner syndrome&Mediterranean macrothrombocytopenia)**
1281. **Ebola virus related diseases**
1282. **Eccrine angiokeratomatous hamartoma**
1283. **Eccrine angiomatous hamartoma EAH**
1284. **Eccrine nevus**
1285. **Eccrine squamous syringometaplasia**
1286. **Ecchymosis**
1287. **Echo virus related diseases**
1288. **Ecthyma simplex= Ulcerative impetigo**
1289. **Ecthyma contagiosum =orf**
1290. **Ecthyma gangrenosum**
1291. **Ectodermal Dysplasia (see next)**
1292. **✓ ADULT: Acro-Dermato-Ungual-Lacrimal-Tooth**
1293. **✓ AEC :Ankyloblepharon, Ectodermal Dysplasia, Cleft lip/palate=Hay–wells syndrome)**
1294. **✓EEC ectodermal Dysplasia, ectrodactyly, cleft lip/palate**
1295. **✓Hidrotic ectodermal Dysplasia=clouston syndrome**
1296. **✓immunodeficiency, myopathy and ED**
1297. **✓ Pure hair and nail ectodermal dysplasia (PHNED)**
1298. **✓Ectodermal Dysplasia nectin 4**
1299. **✓Nectinopathies: cleft lip&palate**
1300. **✓ED, ectrodactyly and macular dystrophy (EEM)**
1301. **✓ Rapp Hodgkin Syndrome RHS**
1302. **✓ Acrorenal field defect, ED, and lipoatrophic diabetes (AREDYLD) syndrome**

1303. ✓Hypohidrotic ED–immune deficiency
1304. ✓Hypohidrotic Ectodermal Dysplasia=christ–siemens–touraine syndrome=X-linked hypohidrotic ectodermal Dysplasia (XLHED)
1305. ✓Autosomal Recessive Hypohidrotic Ectodermal Dysplasia
1306. ✓Autosomal Dominant Hypohidrotic Ectodermal Dysplasia
1307. ✓Hypohidrotic Ectodermal Dysplasia with hypothyroidism and agenesis of the corpus callosum
1308. ✓Hypohidrotic Ectodermal Dysplasia with hypothyroidism and ciliary dyskinesia (hedh syndrome)
1309. ✓Ectodermal Dysplasia-skin fragility syndrome (ED-SFS) = McGrath syndrome
1310. ✓Ectodermal Dysplasia with palatal paralysis
1311. ✓Ectodermal Dysplasia with mental retardation and syndactyly
1312. ✓Ectodermal Dysplasia with syndactyly Syndrome (EDSS
1313. ✓Ectodermal Dysplasia and neurosensory deafness (mikaelian syndrome)
1314. ✓Ectodermal Dysplasia with adrenal cyst (odonto-onycho-hypohidrotic dysplasia with midline scalp defect)
1315. ✓Ectodermal Dysplasia with severe mental retardation
1316. ✓Ectodermal Dysplasia with natal teeth (turnpenny type)
1317. ✓Ectodermal Dysplasia with distinctive facies and preaxial polydactyl
1318. ✓ED with onychogryphosis
1319. ✓Polycystic brain associated with Ectodermal Dysplasia
1320. ✓ED with short stature
1321. ✓lelis syndrome (hypohidrotic Ectodermal Dysplasia with acanthosis nigricans)
1322. ✓Margarita island Ectodermal Dysplasia
1323. ✓Arthrogryposis and ectodermal Dysplasia (tricho-oculo-dermo-vertebral syndrome; alves syndrome)
1324. ✓Mesomelic dwarfism –skeletal abnormalities –Ectodermal Dysplasia
1325. ✓Cranio-Ectodermal Dysplasia syndrome (levin syndrome i; sensenbrenner syndrome

1326. ✓Ectodermal Dysplasia with Alopecia–onychodysplasia–hypohidrosis – deafness
1327. ✓Ectodermal Dysplasia with cataracts and hearing defects
1328. Ectomesenchymal chondromyxoid tumor
1329. Ectomesenchymoma
1330. Ectopic breast tissue
1331. Ectopic cilia
1332. Eczema= contact dermatitis
1333. Airborn contact dermatitis
1334. Allergic contact dermatitis= Allergic eczema (pigmented & lichenoid& erythema multiforme & pigmented purpura& pseudolymphomatous & granulomatous & pustular)
1335. Apron eczema
1336. Asteatotic Eczema = eczema craquelé =winter eczema =winter itch=desiccation dermatitis
1337. Baboon Eczema = systemic allergic contact dermatitis
1338. Car fitter dermatitis=Mechanic hand dermatitis
1339. Car Seat Dermatitis
1340. Circumostomy eczema
1341. Commode dermatitis= Toilet-seat dermatitis
1342. Discoid Eczema =nummular dermatitis=nummular eczema
1343. Oid-oid disease
1344. Disseminated Eczema= autosensitization dermatitis
1345. Plant Dermatitis
1346. Post traumatic eczema
1347. Eczema due to war chemicals
1348. Dyshidrotic eczema =Pompholyx= acute and recurrent vesicular hand dermatitis = vesicular eczema
1349. Eczema: follicular type
1350. Frictional lichenoid dermatitis=Dermatitis papulosa juvenilis=frictional lichenoid eruption=Sutton's summer prurigo=sandbox dermatitis=Dermatitis papulosa adultorum

1351. **Hand eczema (see next)**
1352. **✓Pompholyx (vesicular eczema of palms and soles), dyshidrotic eczema**
1353. **✓Patchy vesiculosquamous eczema**
1354. **✓Hyperkeratotic palmar eczema (tylotic eczema)= (eczema keratoticum)**
1355. **✓Recurrent focal palmar peeling= desquamation en aires**
1356. **✓Discoid eczema (nummular eczema)**
1357. **✓Wear and tear dermatitis (housewives dermatitis)**
1358. **✓Ring eczema**
1359. **✓Finger tip eczema=Pulpitis**
1360. **✓Apron eczema**
1361. **✓Gut=slaughterhouse eczema**
1362. **✓Chronic acral dermatitis**
1363. **Irritant contact dermatitis (ICD) (see next)**
1364. **✓ Acute irritant contact dermatitis**
1365. **✓ Cumulative irritant contact dermatitis**
1366. **✓ Acute delayed irritant contact dermatitis**
1367. **✓ Irritant reaction irritant contact dermatitis**
1368. **✓ Asteatotic dermatitis**
1369. **✓ Traumatic irritant contact dermatitis**
1370. **✓ Pustular and acneiform irritant contact dermatitis**
1371. **✓ Non-erythematous irritant contact dermatitis**
1372. **✓ Subjective or sensory irritant contact dermatitis**
1373. **✓ Contact urticaria**
1374. **✓ Frictional irritant contact dermatitis**
1375. **✓Airborne Irritant Contact Dermatitis**
1376. **Infectious eczematoid dermatitis**
1377. **Lymphomatoid contact dermatitis**
1378. **Microbial eczema of the feet**
1379. **Eczema-like psoriasiform dermatitis**
1380. **Eczema marginatum**
1381. **Eczema herpeticum= kaposi varicelliform eruption= Herpetic eczema**

1382. **Eczema coxsackium**
1383. **Eczema molluscatum**
1384. **Eczema monkeypoxicum**
1385. **Eczema vaccinatum**
1386. **Palmoplantar eczema**
1387. **Slobber dermatitis**
1388. **Stasis dermatitis= gravitational dermatitis = varicose eczema =congestion eczema**
1389. **Systemic contact dermatitis**
1390. **Ehlers–Danlos syndrome =Cutis hyperelastica (see next)**
1391. **✓classic**
1392. **✓classic-like**
1393. **✓cardiac valvular**
1394. **✓Hypermobile**
1395. **✓Vascular**
1396. **✓kyphoscoliotic**
1397. **✓Musculocontractural**
1398. **✓Arthrochalasia**
1399. **✓Dermatosparaxis**
1400. **✓periodontal**
1401. **✓Brittle cornea syndrome**
1402. **✓Myopathic**
1403. **✓Spondylodysplastic**
1404. **✓Fibronectin -deficient eds**
1405. **✓ Tenascin -x deficient)**
1406. **✓Kresse syndrome (ehlers–danlos,progeroid type)**
1407. **✓Hernandez (ehlers–danlos-like) syndrome**
1408. **Elaioconiosis**
1409. **Elastoderma**
1410. **Elastofibroma dorsi**
1411. **Elastoma of dubreuilh**

1412. **Elastoma=elastic tissue nevus=nevus elasticus**
1413. **Elastosis linearis rubra nasi**
1414. **Elastosis perforans serpiginosa**
1415. **Elastotic nodules of the ear**
1416. **Juvenile elastoma**
1417. **Papular elastorrhexis =Papular elastosis**
1418. **Perforating calcific elastosis**
1419. **Elephantiasis nostras verrucosa**
1420. **Elephantiasic pretibial myxedema**
1421. **Elephantiasis neuromatosa**
1422. **Elephantine psoriasis**
1423. **Ellis–van creveld syndrome=chondroectodermal dysplasia**
1424. **Elejalde syndrome**
1425. **Emberger syndrome**
1426. **Emboli**
1427. **Cholestrol Emboli**
1428. **Oxalate Emboli**
1429. **Emboli cutis medicamentosa= Nicolau's syndrome**
1430. **Emergomycosis**
1431. **Emmonsiosis**
1432. **Encephalocele**
1433. **Endocarditis related skin lesions**
1434. **Endocrine: Acromegaly related skin lesions**
1435. **Endocrine: Addison disease related skin lesions**
1436. **Endocrine: Cushing syndromen related skin lesions**
1437. **Endocrine: Hyperthyroidism related skin lesions**
1438. **Endocrine: Hypothyroidism related skin lesions**
1439. **Endocrine: Pheochromocytoma**
1440. **Endocrine mucin-producing sweat gland carcinoma (EMPSGC)**
1441. **Engraftment syndrome**
1442. **Enterobiasis (pinworms)**
1443. **Entomophtoromycosis= Subcutaneous phycomycosis (basidiobolomycosis and rhinoentomophtoromycosis)**

1444. **Envelopathies**
1445. **Eosinophilic annular erythema**
1446. **Eosinophilic dermatosis of hematologic malignancy (EDHM)**
1447. **Eosinophilic fasciitis**
1448. **Eosinophilic granuloma**
1449. **Eosinophilic myalgia syndrome**
1450. **Eosinophilic panniculitis**
1451. **Eosinophilic pustular folliculitis**
1452. **Eosinophilic ulcer tongue= eosinophilic nodule of the tongue**
1453. **Eosinophilic, polymorphic, and pruritic eruption associated with radiotherapy**
1454. **Epidermization of lip**
1455. **Epidermoid cyst= Infundibular cyst=Epidermal cyst=Epidermal inclusion cyst**
1456. **Verrucous epidermoid cysts**
1457. **Epidermodysplasia verruciformis EDV**
1458. **Epidermolysis bullosa acquisita EBA=Acquired epidermolysis bullosa (see next)**
1459. **✓Non-inflammatory mechanobullous disease**
1460. **✓BP-like presentation**
1461. **✓Mucous membrane pemphigoid-like presentation**
1462. **✓Brunsting–Perry cicatricial pemphigoid-like presentation**
1463. **✓linear IgA bullous dermatosis-like presentation**
1464. **Epidermolysis bullosa simplex (EBS) (see next)**
1465. **✓EBS superficialis (ebss)**
1466. **✓Acral peeling skin syndrome**
1467. **✓Acantholytic superficialis (ebs acanth)**
1468. **✓Skin fragility syndromes:**

- **Desmoplakin deficiency (ebs-desmoplakin; skin fragility–wooly hair syndrome)**
- **plakoglobin deficiency (ebs-plakoglobin;skin fragility–plakoglobin deficiency)**

- **plakophilin deficiency (ebs-plakophilin;skin fragility–ectodermal dysplasia syndrome)**

1469. ✓ **EBS due to klhl24 ad kelch-like protein 24**
1470. ✓ **EBS, generalized severe (dowling–meara)= Epidermolysis bullosa herpetiformis**
1471. ✓ **EBS, generalized intermediate**
1472. ✓ **EBS,other generalized ,koebner type**
1473. ✓ **EBS, localized (weber–cockayne)**
1474. ✓ **EBS with mottled pigmentation**
1475. ✓ **EBS with pyloric atresia ar plectin; α 6 β 4 integrin**
1476. ✓ **EBS, AR-BP230 deficiency ar bullous pemphigoid antigen 1 (bp230)**
1477. ✓ **EBS, AR-exophilin 5 deficiency**
1478. ✓ **EBS, AR-KRT14**
1479. ✓ **EBS, migratory circinate**
1480. ✓ **EBS-ogna ad plectin**
1481. ✓ **EBS with muscular dystrophy(EBS-MD)**
1482. ✓ **Lethal acantholytic epidermolysis bullosa**
1483. **Dystrophic Epidermolysis bullousa DEB (see next)**
1484. ✓ **RDEB variants: localized, pretibial, pruriginosa**
1485. ✓ **RDEB, bullous dermolysis of the newborn**
1486. ✓ **RDEB, generalized intermediate (non-Hallopeau-Siemens type)**
1487. ✓ **RDEB, generalized severe (hallopeau–siemens)**
1488. ✓ **RDEB, inversa**
1489. ✓ **RDEB centripetalis**
1490. ✓ **DDEB, bullous dermolysis of the newborn**
1491. ✓ **DDEB, generalized Intermediate DDEB**
1492. ✓ **DDEB variants: acral, pretibial, pruriginosa, nails only**
1493. ✓ **Bart syndrome**
1494. ✓ **Albo-papuloid epidermolysis bullosa nigricans of pasinin**
1495. ✓ **Cockayne-touraine syndrome**

1496. **Junctional Epidermolysis bullousa JEB (see next)**
1497. ✓ **JEB with pyloric atresia**
1498. ✓**carmi syndrome**
1499. ✓**JEB with respiratory and renal involvement ar α 3 integrin subunit**
1500. ✓**JEB miti, generalized intermediate =Non herlitz generalized =Generalized atrophic benign epidermolysis bullosa GABEB= Epidermolysis bullosa atrophicans generalisata mitis**
1501. ✓**JEB, generalized severe Gravis (letalis,Herlitz disease)**
1502. ✓ **JEB, inversa**
1503. ✓ **JEB, late onset**
1504. ✓**JEB, localized**
1505. ✓ **JEB-laryngo-onycho-cutaneous syndrome=shabbir's syndrome**
1506. **Epidermolytic acanthoma**
1507. **Cutaneous epithelioid angiomatous nodule (CEAN)**
1508. **Epithelioid hemangioma**
1509. **Epithelioid hemangioendothelioma**
1510. **Epithelioid cell histiocytoma= Epithelioid fibrous histiocytoma**
1511. **Epithelioid sarcoma**
1512. **Epithelioid sarcoma-like hemangioendothelioma(pseudomyogenic hemangioendothelioma)**
1513. **Epithelioma cuniculatum**
1514. **Episodic angioedema with eosinophilia=Gleich syndrome**

Eponymous signs in dermatology (see next)

1515. ✓**Albright's dimple sign**
1516. ✓**Alcohol wipe sign**
1517. ✓**Anal groove sign**
1518. ✓**Antenna sign**
1519. ✓**Apple jelly sign**
1520. ✓**Asboe-hansen sign**
1521. ✓**Auspitz sign**
1522. ✓**Axillary ecchymosis sign**

1523. ✓**Ball-SITE sign**
1524. ✓**Bare underbelly sign**
1525. ✓**Barnett's sign**
1526. ✓**Battle's sign (mastoid ecchymosis)**
1527. ✓**Beauty and the beast sign**
1528. ✓**Benediction sign**
1529. ✓**Berliner sign**
1530. ✓**Besnier's sign**
1531. ✓**Biederman's sign**
1532. ✓ **Biett's sign**
1533. ✓ **Bolognia sign**
1534. ✓ **Breakfast, lunch, and dinner sign**
1535. ✓**Blink sign**
1536. ✓**Blue dot sign**
1537. ✓**Blue pseudoveil sign**
1538. ✓**Blue underpants' sign**
1539. ✓**Blueberry muffin sign**
1540. ✓**Bonbon toffee sign**
1541. ✓**Borsieri's sign**
1542. ✓**Bote sign**
1543. ✓**Branham's sign (nicoladoni sign)**
1544. ✓**Bravermans sign**
1545. ✓**Brenner sign**
1546. ✓**Brownie nose sign**
1547. ✓**Bull's head sign**
1548. ✓**Burnt matchstick sign**
1549. ✓**Buschke-ollendorff sign**
1550. ✓**Butterfly sign**
1551. ✓**Buttonhole sign**
1552. ✓**Calabar sign**
1553. ✓**Candle wax sign**

1554. ✓Caput medusae
1555. ✓Carpet tack sign (tin tack sign, cat tongue sign)
1556. ✓Casal's necklace sign
1557. ✓Cayenne pepper sign
1558. ✓Central dell sign
1559. ✓Chagas–mazza–romaña's sign
1560. ✓Chandelier's sign
1561. ✓Chik sign
1562. ✓Clown nose sign
1563. ✓Cluster of jewels sign (string of pearls, rosette sign
1564. ✓Color-transition sign
1565. ✓Comby sign
1566. ✓Comet sign
1567. ✓Coral bead sign
1568. ✓Corn flake sign
1569. ✓Coudability sign
1570. ✓Coup d'ongle sign
1571. ✓Crowe's sign
1572. ✓Cuff sign
1573. ✓Cullen's sign
1574. ✓Cumulus sign
1575. ✓Darier's sign
1576. ✓Deck-chair sign
1577. ✓Delmege sign
1578. ✓Delta wing jet with contrail sign
1579. ✓Dew drop on rose petal sign
1580. ✓Diamond necklace sign
1581. ✓Dimple sign (fitzpatrick sign)
1582. ✓Dirty neck sign
1583. ✓Dot-in-circle sign
1584. ✓Double edge sign

1585. ✓**Doughnut sign**
1586. ✓**Drip sign**
1587. ✓**Dubois sign**
1588. ✓**Ear lobe sign**
1589. ✓**Ear sign**
1590. ✓**Eastern pancake sign**
1591. ✓**Elbow sign**
1592. ✓**Enamel paint sign (crazy pavement sign= crackled sign= mosaic sign= flaky paint sign)**
1593. ✓**Eyelid sign**
1594. ✓**Fibula sign**
1595. ✓**Filipovitch's sign**
1596. ✓**Flag sign – clinical**
1597. ✓**Flag sign-histopathological**
1598. ✓**Flambeau sign**
1599. ✓**Forscheimer's sign**
1600. ✓**Fountain sign**
1601. ✓**Fountain of youth sign**
1602. ✓**Frank's sign**
1603. ✓**Friar tuck sign**
1604. ✓**Fried egg sign**
1605. ✓**Frog spawn appearance**
1606. ✓**Froment sign**
1607. ✓**Frosted Glass Sign**
1608. ✓**Giovannini sign**
1609. ✓**Goat eye sign**
1610. ✓**Goldstein's toe sign**
1611. ✓**Golf club set sign**
1612. ✓**Gorlin's sign**
1613. ✓**Gottron's sign**
1614. ✓**Greenhow's sign (vagabond's sign)**

1615. ✓Grey turner's sign
1616. ✓Groove sign of greenblatt
1617. ✓Groove sign
1618. ✓Gulliver's sign
1619. ✓Hair collar sign
1620. ✓Hairy palm sign
1621. ✓Halo sign
1622. ✓Hamburger sign
1623. ✓Handle bar sign
1624. ✓Hang-glider sign
1625. ✓Hanging groin sign
1626. ✓Hanging curtain sign
1627. ✓Hatchcock sign
1628. ✓Headlight sign
1629. ✓Heliotrope sign
1630. ✓Hertoghe's sign (queen anne's sign)
1631. ✓Hiker's feet sign
1632. ✓Hildreth's sign
1633. ✓Hoagland sign
1634. ✓Holster sign of dermatomyositis
1635. ✓Hutchinson's sign (melanoma &herpes zoster)
1636. ✓Hypopyon sign
1637. ✓Iceberg sign
1638. ✓Ice-pack sign
1639. ✓Igneous rock sign
1640. ✓Ingram's sign
1641. ✓Isobar sign
1642. ✓Jacquet's sign
1643. ✓Jellinek's sign (rasin's sign)
1644. ✓Jelly sign
1645. ✓Kaposi-stemmer sign

1646. ✓Keining sign
1647. ✓Kerr's sign
1648. ✓Knife cut sign
1649. ✓Lafora's sign
1650. ✓Leopard skin sign
1651. ✓Leser–trelat sign
1652. ✓liddle's sign
1653. ✓Lightening sign
1654. ✓Linear gingival erythema sign
1655. ✓Little red riding hood sign
1656. ✓Lizard skin sign
1657. ✓Love's sign
1658. ✓Lyon's sign
1659. ✓Mace sign
1660. ✓Maroni's sign
1661. ✓Matchbox sign
1662. ✓Meffert's sign
1663. ✓Micro-hutchinson sign
1664. ✓Milian's ear sign
1665. ✓Milk sign (scarlet milk sign)
1666. ✓Mistletoe sign
1667. ✓Mizutani's sign (round finger pad sign)
1668. ✓Muehrcke's sign
1669. ✓Mushroom cloud sign
1670. ✓Myer sign
1671. ✓Nagayama sign
1672. ✓Nail flag sign
1673. ✓Nasal crease sign
1674. ✓Nazzaro's sign
1675. ✓Neighborhood sign
1676. ✓Nikolskiy's sign

1677. ✓Noodle sign
1678. ✓Nose sign (pavithran's nose sign)
1679. ✓Oil drop sign
1680. ✓Oil spot sign
1681. ✓Osler's sign
1682. ✓Ovoid palatal sign
1683. ✓Oyster blister sign
1684. ✓Paired ear creases of the helix sign
1685. ✓Panda and lambda sign
1686. ✓Pastia's sign
1687. ✓Pathergy sign
1688. ✓Patrick yesudian sign
1689. ✓Pearl necklace sign
1690. ✓Pemberton's sign
1691. ✓Peripilar sign
1692. ✓Pink glow sign
1693. ✓Pink rim sign
1694. ✓Pink sign
1695. ✓Pitaluga's sign
1696. ✓Pluck out sign
1697. ✓Poppyfield bleeding sign
1698. ✓Pore sign
1699. ✓Prayer sign
1700. ✓Premlatha's sign
1701. ✓Promontory sign
1702. ✓Punshi's sign
1703. ✓Pup-tent sign
1704. ✓Raccoon sign
1705. ✓Radial crown sign
1706. ✓Rainbow sign
1707. ✓Red planet sign

1708. ✓**Red, white, and blue sign**
1709. ✓**Regularly bended ribbon sign**
1710. ✓**Reverse namaskar sign**
1711. ✓**Ring scales sign**
1712. ✓**Romana's sign**
1713. ✓**Rope sign**
1714. ✓**Rose spots**
1715. ✓**Rosette sign**
1716. ✓**Rudolph sign**
1717. ✓**Russell's sign**
1718. ✓**Sagrada familia sign, digitations and mirror sign**
1719. ✓**Salute sign**
1720. ✓**Samitz's sign**
1721. ✓**Sandwich sign**
1722. ✓**School chair sign**
1723. ✓**Scratch sign (coup d'ongle sign, besnier's sign)**
1724. ✓**Seat rash sign**
1725. ✓**Setting sun sign**
1726. ✓**Shar-pei sign**
1727. ✓**Shawl sign**
1728. ✓**Shelly's sign**
1729. ✓**Shoulder pad sign**
1730. ✓**Shuster's sign**
1731. ✓**Skin crease sign**
1732. ✓**Snakeskin sign**
1733. ✓**Spade sign**
1734. ✓**St.tropez sign**
1735. ✓**Stafne's sign**
1736. ✓**Starburst sign**
1737. ✓**Starry night sky sign**
1738. ✓**Statue of liberty sign**

1739. ✓Sternberg's thumb sign
1740. ✓Stinson sign
1741. ✓String of pearls sign
1742. ✓Tap sign
1743. ✓Tasleem's water jet sign
1744. ✓Tent sign
1745. ✓Thompson's sign
1746. ✓Thumb sign (steinberg sign)
1747. ✓Thumbprint sign
1748. ✓Toothbrush sign
1749. ✓Toy soldier sign
1750. ✓Translucency sign
1751. ✓Triangular nasal notch sign
1752. ✓Triangular sign
1753. ✓Tripe palms sign
1754. ✓Ugly duckling sign
1755. ✓Umbilical sign
1756. ✓Urschel's sign
1757. ✓V- sign
1758. ✓Vieira sign
1759. ✓Volcano sign
1760. ✓Wake sign
1761. ✓Walker- Murdoch's/Wrist sign
1762. ✓Walzel's sign
1763. ✓Wartenberg's sign
1764. ✓White islands in a sea of red sign
1765. ✓Wimberger's sign
1766. ✓Winterbottom's sign
1767. ✓Wipeout sign
1768. ✓Wobble sign
1769. ✓Wrist sign (walker's sign)

1770. ✓Yellow clod sign
1771. ✓Yoga sign

Clothing-related Eponyms (see next)

1772. ✓Cradle cap
1773. ✓Turban tumors
1774. ✓Woolly hair
1775. ✓Spun glass hair
1776. ✓Beaded/necklace hair
1777. ✓Hidden hair
1778. ✓Hair collar sign
1779. ✓Hair pin vessels
1780. ✓Crown vessels
1781. ✓Lacy striae
1782. ✓Velvety plaques
1783. ✓Moroccan leather
1784. ✓Silk road disease=behcet disease
1785. ✓Harlequin armor
1786. ✓Bathing suit appearance
1787. ✓Bare underbelly sign
1788. ✓Shoulder pad sign
1789. ✓Shawl sign
1790. ✓Mask of pregnancy
1791. ✓Blue grey veil
1792. ✓Buttonhole sign
1793. ✓Gaiter region
1794. ✓Holster sign
1795. ✓Mitten hands/feet
1796. ✓Casal's necklace
1797. ✓Necklace of venus
1798. ✓Epstein pearls
1799. ✓String of pearls sign

1800. ✓Pearly appearance
1801. ✓Amniotic band syndrome
1802. ✓Keratodermic sandals
1803. ✓Beaded sign
1804. ✓Coral beads sign
1805. ✓Signet ring sign
1806. ✓Brass knuckles sign
1807. ✓Safety pin sign
1808. ✓Chaining pattern
1809. ✓Perivascular cuffing
1810. ✓Cuff sign
1811. ✓Rosette sign
1812. ✓Papular-purpuric gloves and socks syndrome

Food Eponyms in Dermatology(see next)

1813. ✓Apple jelly nodules
1814. ✓Bean bag cells
1815. ✓Bean' syndrome
1816. ✓Black berry stomatitis
1817. ✓Blueberry muffin baby
1818. ✓Blueberry muffin lesions
1819. ✓Breakfast, lunch and dinner sign
1820. ✓Café-au-lait macules
1821. ✓Cayenne pepper spots
1822. ✓Champagne bottle leg
1823. ✓Cherry angiomas
1824. ✓Corn flake sign
1825. ✓Curry-Hall syndrome
1826. ✓Curry Jones syndrome
1827. ✓Doughnut sign
1828. ✓Fried egg
1829. ✓Garlic clove fibroma

1830. ✓**GEMSS syndrome**
1831. ✓**Grain itch/Barley itch/straw itch/Mattress itch/Prairie itch/cotton weed dermatitis**
1832. ✓**Honey comb atrophy**
1833. ✓**Mango dermatitis**
1834. ✓**Milian citrine skin**
1835. ✓**Mulberry like erosions**
1836. ✓**Mulberry molars**
1837. ✓**Mushroom deformity**
1838. ✓**Oil acne**
1839. ✓**Oil granuloma**
1840. ✓**Oil spot**
1841. ✓**Onion skin cysts**
1842. ✓**Peau d' orange**
1843. ✓**Port wine stain**
1844. ✓**Red Strawberry tongue**
1845. ✓**Salt and pepper fundus**
1846. ✓**Sandwich sign**
1847. ✓**Sausage finger**
1848. ✓**Spaghetti and meat balls**
1849. ✓**Strawberry gums**
1850. ✓**Strawberry hemangioma**
1851. ✓**Sweet syndrome**
1852. ✓**Swiss cheese effect**
1853. ✓**Toasted skin syndrome**
1854. ✓**Tomato catsup fundus**
1855. ✓**Wafer like scales**
1856. ✓**Watermelon stomach**
1857. ✓**White strawberry tongue**

Eponyms of substance abuse (see next)

1858. ✓**Cannabis arteritis**

1859. ✓Betel chewer's mucosa
1860. ✓Chingari sign
1861. ✓Harlequin nails=quitters' nails
1862. ✓Smokers' palate=smokers' keratosis
1863. ✓ Smoker's tongue
1864. ✓Smokers' melanosis
1865. ✓Smoker's face
1866. ✓Smokers' moustache
1867. ✓Smokers' comedones
1868. ✓Smokers callosity
1869. ✓Snuff dippers lesions
1870. ✓Cocaine bugs
1871. ✓Cocaine-induced midline destructive lesions (cimdl)
1872. ✓Cocaine keratosis
1873. ✓Parrot beak nails
1874. ✓Formication
1875. ✓Gummies
1876. ✓Pseudovasculitis
1877. ✓Levamisole purpura
1878. ✓Matchbox sign
1879. ✓Snorters' warts
1880. ✓Ecstasy pimples
1881. ✓Meth mouth
1882. ✓Pentazocine ulcers
1883. ✓Pentazocine ulcers
1884. ✓High pruritus
1885. ✓Pseudoacanthosis nigricans
1886. ✓Puffy hands syndrome
1887. ✓Railroad tracks
1888. ✓Skin popping
1889. ✓Shooter's patch

1890. ✓Shooting tattoo
1891. ✓Track mark
1892. ✓Torniquet hyperpigmentation
1893. ✓Coma blisters
1894. ✓Glue sniffers rah

Syphilis signs(see next)

1895. ✓ Buschke-ollendorff sign
1896. ✓ Barber-pole sign
1897. ✓ Biederman's sign
1898. ✓Bucket handle sign
1899. ✓ Bulldog jaw sign
1900. ✓ Café-au-lait tint sign
1901. ✓ Crown of venus sign
1902. ✓Dermarquay sign
1903. ✓ Dory flop sign
1904. ✓ Du bois sign
1905. ✓ Elliot sign
1906. ✓ Higoumenaki's sign
1907. ✓ Hutchinson teeth sign
1908. ✓ Krisowski's sign
1909. ✓ Mulberry molars
1910. ✓ Natiform skull sign
1911. ✓ Olympian brow sign
1912. ✓ Omnibus sign
1913. ✓ Opera-glass nose sign
1914. ✓ Parrot's nodes
1915. ✓ Sabre shins (fournier's sign)
1916. ✓Sarcophagus sign
1917. ✓ Seeping sign
1918. ✓ Silex's sign
1919. ✓ Split papules

1920. ✓ Virchow's sign
1921. Epstein pearl
1922. Epstein barr virus - infectious mononucleosis =Glandular Fever= Pfeiffer Glandular Fever= Filatov's Disease
1923. Epstein-Barr Virus-Associated Smooth Muscle Tumors
1924. Epulis fissuratum / Epulides
1925. Epulis granulomatosa / Epulides
1926. Ethylmalonic aciduria
1927. Erosive adenomatosis of the nipple
1928. Erosive papulonodular dermatosis
1929. Erosive pustular dermatosis of the scalp
1930. Erosive pustular dermatosis of the leg
1931. Eruptive hypomelanosis (idiopathic)
1932. Eruptive keratoderma of the hands and feet of rahim
1933. Eruptive pseudoangiomatosis
1934. Eruptive recurrent pigmented telangiectasia
1935. Erysipelas
1936. Erysipeloid=Diamond skin disease
1937. Erythema
1938. Erythema ab igne = Toasted skin syndrome = fire stains
1939. Erythema annulare centrifugum (EAC):superficial &deep
1940. Erythema arthriticum epidemicum
1941. Erythema chronicum migrans of lyme disease
1942. Erythema contusiforme
1943. Erythema dyschromicum perstans =ashy dermatosis
1944. Erythema elevatum diutinum EED
1945. Erythema gyratum atrophican of neonate
1946. Erythema gyratum repen
1947. Erythema gyratum repen like urticarial vasculitis
1948. Erythema gyratum repen like bullous lupus
1949. Erythema gyratum repen like Erythema annulare centrifugum
1950. Erythema gyratum repen like psoriasis

1951. **Erythema induratum (nodular vasculitis =erythema induratum of bazin or bazin disease (tuberculous etiology) & erythema induratum of whitfield (non-tuberculous etiology)**
1952. **Erythema infectiosum: fifth disease:slapped cheek syndrome**
1953. **Erythema marginatum**
1954. **Erythema marginatum-like lesions of hereditary angioedema**
1955. **Erythema multiforme (minor & major)**
1956. **EMPACT syndrome (Erythema multiforme associated with Phenytoin And Cranial radiation Therapy)**
1957. **Erythema nodosum EN**
1958. **Erythema nodosum Like lesion in behcet disease**
1959. **Erythema nodosum leprosum**
1960. **Erythema nodosum migrans**
1961. **Erythema nuchae (salmon patch)**
1962. **Erythema palmare hereditarium**
1963. **Erythema papulosa semicircularis recidivans = erythema papulatum centrifugum**
1964. **Erythema punctatum higuchi (EPH)**
1965. **Erythema rhematicum migrans**
1966. **Concentric centrifugal figurate erythema**
1967. **Palpable migratory arciform erythema**
1968. **Erythema scarlatiniforme desquamativum recidivans**
1969. **Erythema toxicum neonatorum =toxic erythema of the newborn**
1970. **Erythrasma**
1971. **Disciform erythrasma**
1972. **Erythrocyanosis**
1973. **Erythroderma**
1974. **Idiopathic erythroderma**
1975. **Erythrodermic trichophyton**
1976. **Erythrokeratolysis hiemalis =keratolytic winter erythema=oudtshoorn disease**
1977. **Erythroleukoplakia**
1978. **Erythroplakia**

1979. **Erythromelalgia= erythralgia= erythermalgia**
1980. **Erythromelanosis follicularis faciei et colli**
1981. **Erythroplasia of queyrat**
1982. **Erythrose péribuccale of brocq**
1983. **Erythrosis pigmentosa mediofacialis**
1984. **Erasmus syndrome**
1985. **Essential fatty acid deficiency**
1986. **Estrogen dermatitis**
1987. **Ewing's sarcoma= peripheral primitive neuroectodermal tumor/PNET**
1988. **Evans syndrome**
1989. **Exserohilum infection**
1990. **Excited skin syndrome**
1991. **Exercise-induced vasculitis =Golfer vasculitis**
1992. **Exfoliative dermatitis**
1993. **Extramedullary hematopoiesis**
1994. **Extravasation injuries**
1995. **Fabricated/induced illness/Munchhausen syndrome by proxy**
1996. **Fabry disease**
1997. **FACE Syndrome : facial-afro–caribbean- childhood eruption**
1998. **Facial dermal dysplasia**
1999. **Facial discoid dermatosis FDD**
2000. **Facial idiopathic granulomas with regressive evolution FIGURE**
2001. **Facio-auriculovertebral syndrome**
2002. **Facio-cutaneous-skeletal syndrome**
2003. **Facio-digito-genital syndrome**
2004. **Facio-genital popliteal syndrome**

Facies in dermatology (see next)

2005. ✓ **Acrogeric facies**
2006. ✓ **Acromegaloid facies**
2007. ✓ **Adenoid facies**
2008. ✓ **Amiodarone facies**
2009. ✓ **Angelic facies**

2010. ✓ **Antonine facies**
2011. ✓ **Asiatic porcelain doll facies**
2012. ✓ **Atopic facies**
2013. ✓ **Bird-like facies**
2014. ✓ **Bloodhound facies**
2015. ✓ **Bull dog face**
2016. ✓ **Cadaveric facies**
2017. ✓ **Cigarette facies**
2018. ✓ **Coarse facies**
2019. ✓ **Dengue facies**
2020. ✓ **Dog=simian facies**
2021. ✓ **Drawn facies**
2022. ✓ **Dysmorphic facies**
2023. ✓ **Elfin facies**
2024. ✓ **Gargoyles facies**
2025. ✓ **Gaunt facies**
2026. ✓ **Greek helmet facies**
2027. ✓ **Grimace-like facies**
2028. ✓ **Hippocratic facies**
2029. ✓ **Latrodectus facies**
2030. ✓ **Leonine facies**
2031. ✓ **Mask like face**
2032. ✓ **Mickey mouse facies**
2033. ✓ **Mitral face**
2034. ✓ **Mongoloid face=flat facies**
2035. ✓ **Monkey facies**
2036. ✓ **Moon facies**
2037. ✓ **Old man facies**
2038. ✓ **Sad man facies**
2039. ✓ **Slapped cheek facies**
2040. ✓ **Tabetic facies**

2041. ✓ **Triangular face**
2042. ✓ **Whistling facies**
2043. **Failed scalp micropigmentation**
2044. **Familial annular erythema**
2045. **Familial atypical mole syndrome**
2046. **Familial cerebral cavernous malformation**
2047. **Familial cold auto-inflammatory syndrome FCAS**
2048. **Familial cutaneous and mucosal venous malformation**
2049. **Familial dysautonomia**
2050. **Familial dyskeratotic comedon**
2051. **Familial disseminated comedones without dyskeratosis (FDCWD)**
2052. **Familial flame nevi**
2053. **Familial gigantic melanocytosis**
2054. **Familial haemophagocytic lymphohistiocytosis**
2055. **Familial histiocytic dermatoarthritis**
2056. **Familial mediterranean fever (FMF)**
2057. **Familial multiple acral mucinous fibrokeratomas**
2058. **Familial multiple café-au-lait spots**
2059. **Familial multiple discoid fibromas (FMDF)= Familial Multiple Trichodiscomas**
2060. **Familial multiple lipomatosis**
2061. **Familial primary cutaneous amyloidosis**
2062. **Familial progressive hyperpigmentation**
2063. **Familial progressive hypo-and hyperpigmentation**
2064. **Familial speckled acral hypopigmentation**
2065. **Factor v leiden mutation**
2066. **Fanconi anemia**
2067. **Far Eastern spotted fever**
2068. **Farber lipogranulomatosis disease= ceramidase deficiency**
2069. **Fascioliasis**
2070. **Fat-storing hamartoma of dermal dendrocytes**
2071. **Faun tail= Lumbosacral Hypertrichosis**
2072. **Fegeler syndrome(acquired capillary malformation)**

2073. **Felty's syndrome**
2074. **Female genital mutilation**
2075. **Ferguson–smith syndrome**
2076. **Ferro-Cerebro-Cutaneous (FCC) syndrome = Neurodevelopmental Disorder with Epilepsy and Hemochromatosis (NEDEPH)**
2077. **Fetal alcohol syndrome**
2078. **Fetal hydantoin syndrome**
2079. **FG syndrome =Opitz-Kaveggia syndrome**
2080. **Fiberglass dermatitis**
2081. **Fibroadenoma**
2082. **Fibro adipose vascular anomaly FAVA**
2083. **Fibroblastic connective tissue nevus**
2084. **fibroblastic rheumatism**
2085. **Fibroelastolytic papulosis of neck**
2086. **Fibroblastoma**
2087. **Desmoplastic Fibroblastoma =Collagenous Fibroma**
2088. **Dermal desmoplastic fibroblastoma**
2089. **Fibroepithelial polyp**
2090. **Fibroepithelioma of pinkus**
2091. **Fibrolipoma**
2092. **Liposurfomas(Acquired fibrolipomas)**
2093. **Fibrolipomatous hamartoma**
2094. **Hemosiderotic fibrolipomatous tumor**
2095. **Fibrofolliculoma**
2096. **Fibrokeratoma**
2097. **Fibroma**
2098. **Fibroma of oral mucous membrane =bite fibroma**
2099. **Fibroma fissuratum**
2100. **Fibromatosis colli**
2101. **Fibromatoses (see next)**
2102. ✓ **Desmoid tumor**
2103. ✓ **knuckle pads (holoderma)**

2104. ✓ pachydermodactyly
2105. ✓ palmar fibromatosis (dupuytren disease)
2106. ✓ penile fibromatosis (peyronie disease)
2107. ✓ plantar fibromatosis (ledderhose disease)
2108. ✓ superficial fascial fibromatosis
2109. Fibrodysplasia ossificans progressive
2110. Fibroelastolytic papulosis
2111. Fibroma of tendon sheath
2112. Fibro-osseous pseudotumor of the digits
2113. Fibrosarcoma **(see next)**
2114. ✓ Classic fibrosarcoma
2115. ✓ Fibromyxoid fibrosarcoma
2116. ✓ Myxofibrosarcoma&malignant fibrous histiocytoma
2117. ✓ Sclerosing epithelioid fibrosarcoma
2118. ✓ Congenital=Infantile Fibrosarcoma
2119. ✓ Adult-type fibrosarcoma
2120. Fibrous hamartoma of infancy
2121. Fibrous papule of nose
2122. Fibrous papule of lips
2123. Fiddler's neck
2124. Finlay–Marks syndrome, scalp–ear–nipple syndrome
2125. Finger pebbles
2126. Fish odour syndrome
2127. Fisher-volavsek syndrome
2128. Fisch syndrome
2129. Fitzsimmons-McLachlan-Gilbert syndrome
2130. Fixed drug eruption FDE
2131. FDE variants (Nonpigmenting&Inverse &Linear& Wandering&Chronic& Cellulitis like& Psoriasiform& Neutrophilic&Erythema dyschromicum perstans like&FDE with pigmentary loss&generalized bullous &linear&Paronychia)
2132. Fixed eczematous eruption

2133. **Fixed food eruption**
2134. **Fixed sunlight eruption**

Flagellate dermatoses (see next)

2135. ✓ **adult-onset still's disease**
2136. ✓ **bendamustine**
2137. ✓ **bleomycin**
2138. ✓ **chikungunya fever induced flagellate pigmentation**
2139. ✓ **child & partner &elder abuse**
2140. ✓ **cnidarian (portuguese man-of-war and jellyfish) stings**
2141. ✓ **dermatitis artefacta**
2142. ✓ **dermatographism**
2143. ✓ **dermatomyositis**
2144. ✓ **docetaxel**
2145. ✓ **excoriations by pruritic conditions**
2146. ✓ **hypereosinophilic syndrome**
2147. ✓ **idiopathic flagellate pigmentation**
2148. ✓ **paederus and other insects**
2149. ✓ **peplomycin**
2150. ✓ **phytophotodermatitis**
2151. ✓ **poison ivy dermatitis**
2152. ✓ **religious punishment**
2153. ✓ **sexual pleasure (sadomasochism)**
2154. ✓ **shiitake mushroom ingestion (raw or undercooked)**
2155. ✓ **torture**
2156. **Flagellate hyperpigmentation**
2157. **Flagellate hypopigmentation**
2158. **Flag-like hypomelanosis**
2159. **Flag-like hypermelanosis**
2160. **Flag-like lentiginosis (including partial unilateral lentiginosis)**
2161. **Flea-borne spotted fever**
2162. **Flea-borne typhus=California flea rickettsiosis**

2163. **Flegel's diseases= hyperkeratosis lenticularis perstans**
2164. **Flinders Island spotted fever**
2165. **Floppy eyelid syndrome**
2166. **Flotch syndrome**
2167. **Florid papillomatosis**
2168. **Florid reactive periostitis**
2169. **Flushing**
2170. **Flynn-aird syndrome**
2171. **Focal facial dermal dysplasia (FFDD)**
2172. **✓ ffdd1 – brauer**
2173. **✓ ffdd2 – brauer–setleis**
2174. **✓ ffdd3 – setleis**
2175. **✓ ffdd4 – preauricular**
2176. **Focal epithelial hyperplasia**
2177. **Focal Fibrous Hyperplasia**
2178. **Folliculosebaceous cystic hamartoma (FSCH)= sebofolliculoma**
2179. **Follicular keratosis**
2180. **Follicular occlusion triad**
2181. **Follicular occlusion tetrade**
2182. **Actinic folliculitis**
2183. **Candidal folliculitis**
2184. **Demodex folliculitis**
2185. **Dissecting terminal hair folliculitis**
2186. **Disseminate and recurrent infundibulofolliculitis (DRIF)= hitch and lund disease**
2187. **Eosinophilic pustular folliculitis infancy**
2188. **Eosonophilic folliculitis of ofuji**
2189. **Eosinophilic folliculitis associated with HIV infection**
2190. **Immunosuppression-associated eosinophilic pustular folliculitis**
2191. **Eosinophilic pustular folliculitis**
2192. **Folliculitis decalvans = Quinquaud's Disease= Acne Decalvans**
2193. **Folliculitis nares perforans**

2194. **Friction folliculitis**
2195. **Gram negative folliculitis**
2196. **Hemorrhagic folliculitis in scurvy**
2197. **Herpes simplex folliculitis**
2198. **Hot tub folliculitis**
2199. **Irritation folliculitis**
2200. **Perforating folliculitis**
2201. **Pityrosporum folliculitis= Malassezia folliculitis**
2202. **Pseudolymphoma folliculitis**
2203. **Folliculitis spinulosa decalvans**
2204. **Sterile neutrophilic folliculitis with perifollicular vasculopathy**
2205. **Superficial pustular folliculitis=Impetigo of bochart**
2206. **Traumatic anserine folliculosis**
2207. **Trunk folliculitis**
2208. **Folliculocystic and collagenous hamartoma**
2209. **Fordyce spots =Ectopic sebaceous glands**
2210. **Foreign body granuloma**
2211. **Formication**
2212. **Fox-fordyce disease =apocrine miliaria**
2213. **Fournier gangrene**
2214. **François syndrome**
2215. **Frank sign (diagonal earlobe crease)**
2216. **Frank ter haar syndrome**
2217. **Franceschetti-klein syndrome= mandibulofacial dysostosis**
2218. **Freckles = Ephelid**
2219. **Frey syndrome**
2220. **Fryns syndrome**
2221. **Friction amyloidosis**
2222. **Frictional asymptomatic darkening of the extensor surfaces (FADES)**
2223. **Frictional hypermelanosis**
2224. **Frictional Keratosis**
2225. **Frictional lichenoid dermatitis**
2226. **Frictional dermal melanosis=lifa disease**

2227. **Frictional sweat dermatitis**
2228. **Fried tooth and nail syndrome**
2229. **Frontometaphyseal dysplasia**
2230. **Frost bite**
2231. **Fucosidosis**
2232. **Fuhrmann syndrome**
2233. **Furuncle**
2234. **Fusariosis=hyalohyphomycosis**
2235. **Futcher's lines**
2236. **Galli-galli diseas**
2237. **Gamasoidosis**
2238. **Galactosialidosis**
2239. **Ganastomiasis**
2240. **Ganglion cyst**
2241. **Gangliosidosis**
2242. **Ganglioneuroma**
2243. **Gangrene**
2244. **Gapo syndrome (growth retardation, alopecia, pseudoanodontia and optic atrophy)**
2245. **Gardner syndrome= familial polyposis of the colon**
2246. **Gardner-associated fibroma**
2247. **Gardner-diamond syndrome=autoerythrocyte sensitization**
2248. **Gas gangrene**
2249. **Gastrocutaneous syndrome**
2250. **Gata2 deficiency/ MonoMAC syndrome**
2251. **Gaucher disease/Sphingolipidoses**
2252. **Gazali-temple syndrome**
2253. **Gemcitabine-induced pseudocellulitis**
2254. **GEMSS Syndrome(glaucoma, lens ectopia, microspherophakia, joint stiffness and stocky short "pseudoathletic" build)**
2255. **Genralized essential telangiectasia**
2256. **Generalized verrucosis (GV)**
2257. **Genodermatoses**

2258. **Geographical flushing of the children's face**
2259. **Geotrichosis**
2260. **Geroderma osteodysplastica**
2261. **Gianotti-crosti syndrome =papular acrodermatitis of childhood= infantile papular acrodermatitis=papulovesicular acrolocated syndrome**
2262. **Giant axonal neuropathy with curly hair**
2263. **Giant cell arteritis =temporal arteritis**
2264. **Giant cell angioblastoma**
2265. **Giant cell fibroma**
2266. **Giant cell granuloma**
2267. **Giant cell lichenoid dermatitis**
2268. **Giant cell tumor tendon sheath= Tenosynovial Giant Cell Tumor**
2269. **Giant Cell Tumor of Soft Tissue**
2270. **Gingival cyst of the adult**
2271. **Gingival cyst of the newborn**
2272. **Gingival fibromatosis**
2273. **Gigantic melanocytosis**
2274. **Gigantomastia**
2275. **Gingival fibromatosis and hypertrichosis**
2276. **Gingival fibromatosis –sparse hair –malposition of teeth**
2277. **Gingival hyperplasia**
2278. **Drug-induced gingival overgrowth**
2279. **Desquamative Gingivitis**
2280. **Necrotizing ulcerative gingivitis**
2281. **Glanders**
2282. **Glandular cheilitis**
2283. **Glanzmann–riniker syndrome**
2284. **Gleich syndrome**
2285. **Nasal Glioma=Nasal cerebral heterotopia**
2286. **Glomeruloid hemangioma**
2287. **Glomangiopericytoma**
2288. **Glomus tumour**
2289. **Glomangioma**

2290. **Glomangiomyoma**
2291. **Glomangiomatosis**
2292. **Glomangiosarcoma**
2293. **Glomuvenous malformation**
2294. **Glossitis**
2295. **Atrophic glossitis**
2296. **Hunter glossitis**
2297. **Median rhomboid glossitis=central papillary atrophy**
2298. **Glossodynia**
2299. **Glucagonoma syndrome**
2300. **Glutaric aciduria**
2301. **Glycogen storage disease**
2302. **Gnas mutation disorders**
2303. **Gnathostomiasis=nodular migratory eosinophilic panniculitis**
2304. **Gnathophyma**
2305. **Gobello Naevus syndrome**
2306. **Goeminne syndrome**
2307. **Goitre**
2308. **Goldenhar syndrome=Oculo-auriculo-Vertebral Dysplasia**
2309. **Goltz -Gorlin syndrome=focal dermal hypoplasia= Goltz syndrome**
2310. **Gomez-lopez-hernandez syndrome= cerebello-trigemino-dermal**
2311. **Gonorrhoea**
2312. **Gonococcemia**
2313. **Gordonia bronchialis**
2314. **Gorlin-goltz syndrome**
2315. **Gorlin–chaudry–moss syndrome**
2316. **Good syndrome**
2317. **Gorham-stout syndrome**
2318. **Goodpasture syndrome**
2319. **✓Gottron papule**
2320. **✓Inverse gottron papule**
2321. **✓Sclerodermoid amyloidosum gottron**

2322. ✓Gottron sign
2323. ✓Gottron-like papule
2324. ✓Gottron acrogeria (gottron syndrome)
2325. ✓Papillomatosis cutis carcinoides of gottron
2326. ✓Darrier-gottron erythrokeratoderma=progressive symmetric erythrokeratoderma
2327. Gout = Arthritis urica &tophi
2328. Gouty panniculitis
2329. Pseudo gout
2330. Granddad syndrome
2331. Graft versus host disease GVHD (acute&chronic)
2332. Graham-little-piccardi-lasseur syndrome
2333. Grönblad-strandberg syndrome
2334. Granular cell tumour
2335. Granular parakeratosis GP **(see next)**
2336. ✓Follicular granular parakeratosis
2337. ✓Granular parakeratosis of eccrine ostia
2338. ✓ Granular parakeratotic acanthoma
2339. ✓Incidental gp
2340. ✓ Intertriginous gp
2341. Granular parakeratotic acanthoma
2342. Granulosis rubra nasi
2343. Granulocytic spongiotic papulovesiculosis
2344. Granuloma annulare **(see next)**
2345. ✓Localized
2346. ✓Generalized
2347. ✓Perforating
2348. ✓Subcutaneous=deep
2349. ✓Disseminated
2350. ✓Patch
2351. ✓ Photodistribute

2352. ✓Follicular pustular variant
2353. ✓Targetoid
2354. ✓Giant
2355. ✓Arcuate dermal erythema
2356. ✓Anetodermic
2357. ✓Acute-onset painful acral
2358. ✓Interstitial
2359. ✓papular
2360. ✓linear
2361. Granuloma faciale
2362. Extrafacial granuloma faciale
2363. Granuloma fissuratum
2364. Granuloma gluteal infantum
2365. Granuloma inguinale
2366. Granuloma multiform
2367. Granulomatous cheilitis
2368. Granulomatous slack skin
2369. Granulomatous vasculitis
2370. Gregg syndrome
2371. Greither's syndrome
2372. Grey baby syndrome
2373. Griscelli –prunieras syndrome (3 types)
2374. Grover disease= transient acantholytic dermatosis
2375. Grybowski type generalized eruptive keratoacanthoma
2376. Guinea worm disease
2377. Gynecomastia
2378. H syndrome
2379. Haber's syndrome
2380. Haberland syndrome
2381. Hajdu-cheney syndrome
2382. Haim-munk syndrome
2383. Hair-thread tourniquet syndrome (HTTS)

2384. **Halo nevus**
2385. **HAIR-AN syndrome hyperandrogenemia, insulin resistance, acanthosis nigricans**
2386. **Haemangioma = Naevus Vasculosus=angiomatous Nevi**
2387. **Diffuse Neonatal Hemangiomatosis =miliary Hemangiomatosis =Disseminated Hemangiomatosis**
2388. **PHACES syndrome (p = posterior fossa (this refers to possible abnormal structures in the brain, particularly the cerebellum.)&h = hemangioma&a = arterial (this refers to possible abnormal arteries in the brain, neck or chest.)&c = cardiac (this refers to possible heart defects or abnormalities of the large blood vessels connected to the heart.)&e = eyes (this refers to possible eye or vision issues&s = sternal cleft)**
2389. **LUMBAR syndrome (lower-body hemangioma& urogenital abnormalities=ulceration&myelopathy (severe compression) of the spinal cord&bony deformities&anorectal malformations and arterial anomalies& renal anomalies)**
2390. **PELVIS syndrome (perineal haemangioma&external genitalia malformations & lipomyelomeningocele&vesicorenal abnormalities&imperforate anus& skin tag)**
2391. **Noninvoluting congenital haemangioma NICH**
2392. **Rapidly involuting congenital haemangioma RICH**
2393. **Partially involuting congenital haemangioma PICH**
2394. **Multifocal infantile haemangiomas**
2395. **Haemangiopericytoma**
2396. **Haemochromatosis**
2397. **Hailey - Hailey disease= familial benign chronic pemphigus**
2398. **Hair cast**
2399. **Hair-follicle nevus**
2400. **Hair heterochromia**
2401. **Hair matrix cyst**
2402. **Red hair=rutilism**
2403. **Hair transplant related diseases**
2404. **Hair Weathering**

2405. **Hair Photoaging**
2406. **Halo dermatitis**
2407. **Halo scalp ring**
2408. **Halogenoderma**
2409. **Hallermann–streiff syndrome=françois dyscephalic syndrome=Oculomandibulodyscephaly with Hypotrichosis**
2410. **Hand-foot-and-mouth disease HFMD**
2411. **Pseudomonas hand foot syndrome**
2412. **Hand foot skin reaction**
2413. **Hand foot skin syndrome**
2414. **Hand–foot–genital syndrome**
2415. **Haploinsufficiency of A20**
2416. **Happle–tinschert syndrome**
2417. **Harlequin colour change of newborn**
2418. **Harlequin syndrome**
2419. **Hartnup disease**
2420. **Hay-wells syndrome**
2421. **Hayden syndrome**
2422. **Heamatohidrosis**
2423. **Heamngiopericytoma**
2424. **Hemolytic uremic syndrome**
2425. **Heart hand syndrome**
2426. **Heat cramps**
2427. **Heat edema**
2428. **Heat stroke**
2429. **Heat syncope**
2430. **Heat-related illnesses**
2431. **Heberden nodule**
2432. **Heck disease=focal epithelial hyperplasia**
2433. **Black heel =talon noir-=calcaneal petechiae**
2434. **Heel-line hyperpigmentation**
2435. **Heel fissures**
2436. **Heel stick injury**

2437. **Heel stick calcinosis**
2438. **Hematoma**
2439. **Hemifacial atrophy**
2440. **Hemifacial mixed appendageal tumor**
2441. **Hemihyperplasia lipomatosis**
2442. **Hemi-maxillo-facial dysplasia**
2443. **Hemochromatosis**
2444. **Hemorrhagic insect bites**
2445. **Hemosiderotic fibrolipomatous tumor (HFLT)**
2446. **Hemophagocytic syndrome**
2447. **Henna allergic dermatitis**
2448. **Hennekam lymphangiectasia–lymphedema syndrome**
2449. **Henoch schonlein purpura= anaphylactoid purpura**
2450. **Heparin-induced thrombocytopenia syndrome (HITS)=Heparin-associated thrombocytopenia with thrombosis (HATT) syndrome**
2451. **He remained (Hematologic related malignancy induced eosinophilic dermatoses)**
2452. **Hereditary acrokeratotic poikiloderma**
2453. **Hereditary alpha tryptasemia**
2454. **Hereditary angioedema**
2455. **Hereditary benign intraepithelial dyskeratosis**
2456. **Hereditary benign telangiectasia**
2457. **Hereditary congenital hypopigmented and hyperpigmented macules (westerhof syndrome)**
2458. **Hereditary fibrosing poikiloderma**
2459. **Hereditary generalized punctate leukoderma**
2460. **Hereditary gingival fibromatosis**
2461. **Hereditary hemorrhagic telangiectasia =osler-weber-rendu syndrome**
2462. **Hereditary leiomyomatosis and renal cell cancer (HLRCC)**
2463. **Hereditary mucoepithelial dysplasia**
2464. **Hereditary multiple exostosis (diaphyseal aclasis)**
2465. **Hereditary multiple lipomas**
2466. **Hereditary neurocutaneous angiomatosis**

2467. **Hereditary nonsyndromic multiple basal cell carcinoma**
2468. **Hereditary onycho-osteodysplasia (HOOD)**
2469. **Hereditary osteo-onychodysplasia**
2470. **Hereditary papulotranslucent acrokeratoderma**
2471. **Hereditary premature canities**
2472. **Hereditary sclerosing poikiloderma of weary**
2473. **Hereditary acrokeratotic poikiloderma =Weary syndrome=Dowling syndrome**
2474. **Hereditary sensory and autonomic neuropathies (HSAN):(IA&IB&IC&II&III&IV& V&HSAN with spastic paraplegia)**
2475. **Heerfordt-Waldenström syndrome**
2476. **Hermansky pudlak syndrome (9 types)**
2477. **Herpangina**
2478. **Herpes simplex (orolabial& gingivostomatitis & genital & gladiatorum & folliculitis & encephalitis & proctitis & whitlow)**
2479. **Herpes simplex (neonatal& intrauterine)**
2480. **Herpetic geometric glossitis**
2481. **Herpes vegetans**
2482. **Herpes zoster=shingles (SEE next)**
2483. **✓Classic**
2484. **✓Abortive**
2485. **✓Hemorrhagic**
2486. **✓Bullous**
2487. **✓Gangrenous**
2488. **✓Disseminated**
2489. **✓Zoster sine herpes**
2490. **✓ Duplex symmetricus**
2491. **✓Duplex bilateralis**
2492. **✓Duplex unilateralis**
2493. **✓Herpetic zoster folliculitis**
2494. **Heterochromia**
2495. **Heterotopic neural tissue of the skin**

2496. **Heterotopic salivary gland tissue**
2497. **Hibernoma**
2498. **Hidroacanthoma simplex=Intraepidermal Poroma**
2499. **Malignant hidroacanthoma simplex**
2500. **Neutrophilic eccrine hidradenitis NEH**
2501. **Idiopathic/recurrent palmoplantar hidradenitis**
2502. **Hidradenitis suppurativa HS=acne inversa=verneuil's disease**
2503. **Hidradenoma**
2504. **Apocrine hidradenoma**
2505. **Hidradenoma papilliferum**
2506. **Poroid Hidradenoma**
2507. **Hidradnocarcinoma**
2508. **Hidradenocarcinoma papilliferum**
2509. **Hidrocystoma (apocrine& eccrine)**
2510. **Solitary (smith) hidrocystoma**
2511. **Hirsutism**

Histiocytosis

2512. **LCH langerhan cell histiocytosis(see next)**
2513. **Adult type langerhan cell histiocytosis**
2514. ✓ **letterer-siwe disease**
2515. ✓ **Hand-schüller-christian disease**
2516. ✓ **Congenital self-healing reticulohistiocytosis (CSHR) =hashimoto-pritzker disease**
2517. ✓ **Eosinophilic granuloma**
2518. **Non LCH langerhan cell histiocytosis =Non-X histiocytosis=Class II histiocytosis (see next)**
2519. ✓ **Progressive nodular histiocytosis PNH= Spindle Cell Xanthogranuloma**
2520. ✓ **Hereditary Progresive mucinosis histiocytosis**
2521. ✓ **Papular xanthoma**
2522. ✓ **Generalized eruptive histiocytosis / histiocytoma GEH**
2523. ✓ **Benign cephalic histiocytosis= Papular Histiocytoses of The Head**

2524. ✓ **Indeterminate histiocytosis**
2525. ✓ **Giant cell reticulohistiocytoma**
2526. ✓ **Solitary reticulohistiocytoma= Solitary Epithelioid Histiocytoma**
2527. ✓ **Adult xanthogranuloma**
2528. ✓ **Juvenile xanthogranuloma**
2529. ✓ **Necrobiotic xanthogranuloma**
2530. ✓ **Xanthoma disseminatum=Montgomery syndrome=Disseminated xanthosiderohistiocytosis**
2531. ✓ **Multicentric reticulohistiocytosis**
2532. ✓ **Erdheim-chester disease= Uber Lipoid Granulomatosis)**
2533. ✓ **Kikuchi-Fujimoto Disease= histiocytic necrotizing lymphadenitis**
2534. ✓ **Familial sea blue histiocytosis**
2535. ✓ **Hemophagocytic lymphohistiocytosis**
2536. ✓ **Rosai–dorfman disease (sinus histiocytosis)**
2537. **Malignant Histiocytosis=Class III histiocytosis (see next)**
2538. ✓ **Histiocytic sarcoma**
2539. ✓ **Langerhans cell sarcoma**
2540. ✓ **Follicular dendritic cell sarcoma**
2541. ✓ **Interdigitating dendritic cell sarcoma**
2542. ✓ **True histiocytic lymphoma= reticulum cell sarcoma=histiosarcoma**
2543. **crystal-storing histiocytosis**
2544. **Histoplasmosis= Darling's Disease= Ohio Valley Disease**
2545. **Hitch syndrome (hearing impairment, undescended testis, circumferential skin creases, and mental handicap)**
2546. **Hobnail hemangioma=targetoid hemosiderotic lymphatic malformation=Targetoid Hemosiderotic Hemangioma= Hobnail Lymphatic Malformation**
2547. **Hobnail hemangioendothelioma family (retiform hemangioendothelioma and papillary intralymphatic angioendothelioma)**
2548. **Holocarboxylase synthetase deficiency**

2549. **Holt-oram syndrome**
2550. **Homocystinuria**
2551. **Hooft disease**
2552. **Hookworm disease**
2553. **Hope disease**
2554. **HOPP (hypotrichosis, acro-osteolysis=onychogryphosis, palmoplantar keratoderma, and periodontitis) syndrome**
2555. **Horizontal forehead lines**
2556. **Hornstein-knickenberg syndrome**
2557. **Hoyeraal–hreidarrson syndrome**
2558. **Human monocytotropic ehrlichioses**
2559. **Human granulocytic anaplasmosis**
2560. **Ehrlichia muris-like agent ehrlichiosis**
2561. **Hunan hand syndrome**
2562. **Huntley's papule**
2563. **✓Hutchinson's sign**
2564. **✓Hutchinson's triad**
2565. **✓Hutchinson's teeth**
2566. **✓Hutchinson melanotic freckle**
2567. **✓Hutchinson's melanotic disease**
2568. **✓Hutchinson–gilford progeria syndrome**
2569. **✓Hutchinson's facies**
2570. **✓Hutchinson's angina**
2571. **✓Hutchinson–horton syndrome**
2572. **✓Hutchinson's prurigo**
2573. **✓Hutchinson's patch**
2574. **✓Hutchinson's dyshidrosis**
2575. **Hyalohyphomycosis**
2576. **Hybrid cysts**
2577. **Hydatid cyst/Echinococcosis**
2578. **Hydroa vacciniforme lymphoproliferative disorder**
2579. **Hydroa vacciniforme=Bazin's hydroa vacciniforme**

2580. **Hydroa vacciniforme-like cutaneous t-cell lymphoma**
2581. **Hydroxyurea-induced ulcers**
2582. **4-hydroxybutyric aciduria**
2583. **Hyper eosinophilic syndrome HES**
2584. **Hypereosinophilic dermatitis of nir-westfried**
2585. **Hyperirritable skin**
2586. **Hyper IgD syndrome**
2587. **Hyper IgE syndrome=job syndrome HIES**
2588. **Hyper linearity palm -sole**
2589. **Hyperkeratotic flexural erythema**
2590. **Hyperkeratotic cutaneous capillary–venous malformation**
2591. **Hypertrophic scar**
2592. **Hyperhidrosis**
2593. **Hyperpigmented macules on the face of young children (HMFYC)**
2594. **Hypertrichosis (see next)**
2595. **✓Congenital hypertrichosis lanuginose**
2596. **✓X-linked hypertrichosis (congenital generalized hypertrichosis)**
2597. **✓Acquired generalized hypertrichosis**
2598. **✓Acquired hypertrichosis lanuginosa**
2599. **✓universal hypertrichosis**
2600. **✓Ambras syndrome**
2601. **✓x-linked hypertrichosis**
2602. **✓Hypertrichosis of the auricle**
2603. **✓Hypertrichosis of the nasal tip**
2604. **✓Trichomegaly of the eyelashes**
2605. **✓Hypertrichosis of the eyebrows**
2606. **✓prepubertal hypertrichosis**
2607. **✓Hypertrichosis cubiti**
2608. **✓Anterior cervical hypertrichosis**
2609. **✓Posterior cervical hypertrichosis**
2610. **✓Hairy palms and soles**

2611. ✓Hairy ear pinna
2612. ✓Hairy elbows syndrome
2613. ✓Gingival fibromatosis with hypertrichosis
2614. ✓Acromegaly with hypertrichosis
2615. Hypertrophy of circumvallate papillae
2616. Hypertrophy of foliate papillae
2617. Hypertrophy of the fungiform papillae
2618. Hypervitaminosis A
2619. Hypervitaminosis B (3,6)
2620. Hypervitaminosis D
2621. Hypervitaminosis E
2622. Hypodermatitis sclerodermaformis
2623. Hypodontia and nail dysgenesis
2624. Hypoglossia-hypodactylia anomaly
2625. Hypohidrosis
2626. Hypohidrosis with diabetes insipidus= fleck's syndrome
2627. Hypohidrosis with neurolabyrinthitis
2628. Hypomelia–hypotrichosis–facial
2629. Hypophosphatasia (HPP)
2630. Hypopigmented scar
2631. Hypothenar hammer syndrome
2632. **Hypotrichosis (see next)**
2633. ✓ Hypotrichosis, marie
2634. ✓ Hypotrichosis, marie unna
2635. ✓ Hypotrichosis simplex
2636. ✓ Hypotrichosis simplex of the scalp
2637. ✓ Hypotrichosis with juvenile macular dystrophy
2638. ✓ Hypotrichosis with recurrent skin "vesicles"
2639. ✓ Localized autosomal recessive hypotrichosis
2640. ✓ Hypotrichosis–lymphedema–telangiectasia syndrome

2641. **Ichthyosis (see next)= Fish-skin disease=alligator skin**
2642. ✓**Ichthyosis vulgaris=ichthyosis simplex**
2643. ✓ **Lamellar Ichthyosis =lamellar exfoliation of the newborn**
2644. ✓ **Autosomal dominant Lamellar Ichthyosis**
2645. ✓ **X linked recessive Ichthyosis = steroid sulfatase deficiency**
2646. ✓**Collodion baby/ Congenital self-healing collodion baby**
2647. ✓**Harlequin ichthyosis = ichthyosis congenita gravior =Harlequin fetus**
2648. ✓**Bullous congenital ichthyosiform erythroderma BCIE=Epidermolytic ichthyosis of brocq=Bullous Ichthyosis**
2649. ✓**Cyclic ichthyosis with epidermolytic hyperkeratosis**
2650. ✓**Congenital ichthyosiform erythroderma= non-bullous congenital ichthyosiform erythroderma NBCIE= Ichthyosis Congenita Type 1**
2651. ✓**Annular epidermolytic ichthyosis**
2652. ✓**Mosaic epidermolytic ichthyosis=epidermolytic type of epidermal nevus**
2653. ✓**Autosomal recessive exfoliative ichthyosis**
2654. ✓**Autosomal recessive congenital ichthyosis ARCI**
2655. ✓**Autosomal recessive ichthyosis with hypotrichosis (ARIH)**
2656. ✓**Autosomal recessive keratoderma-ichthyosis-deafness syndrome ARKID**
2657. ✓**Chanarin–dorfman syndrome =Neutral lipid storage disease with ichthyosis**
2658. ✓**CHILD syndrome: congenital hemidysplasia with ichthyosiform erythroderma and limb defects**
2659. **CHIME(Colobomas, Heart defect, Ichthyosiform dermatosis, Mental retardation, Ear anomalies)= Zunich neuroectodermal syndrome**
2660. ✓**Comel -Netherton syndrome= Ichthyosis linearis circumflexa**
2661. ✓**Conradi–hünermann–happle syndrome= X-linked dominant chondrodysplasia punctata type 2**
2662. **Rhizomelic chondrodysplasia punctate (autosomal recessive chondrodysplasia punctata type 1)**
2663. ✓ **Bathing suit Ichthyosis**

2664. ✓Ichthyosis bullosa of siemens =superficial epidermolytic ichthyosis=Ichthyosis exfoliativa
2665. ✓Ichthyosis cribriformis
2666. ✓Ichthyosis en confetti=Confetti Ichthyosis=Congenital Reticular Ichthyosiform Erythroderma CRIE =Ichthyosis with Confetti= Ichthyosis Variegata
2667. ✓Ichthyosis follicularis with atrichia and photophobia IFAP
2668. ✓Ichthyosis hystrix= ichthyosis hystrix curth-macklin=porcupine man=systematized verrucous nevus
2669. ✓HID: hystrix-like ichthyosis with deafnes
2670. ✓Ichthyosis prematurity syndrome IPS
2671. ✓Ichthyosis with hypogonadism
2672. ✓Ichthyosis, spastic quadriplegia, mental retardation ISQMR
2673. ✓Ichthyosis-hypotrichosis-sclerosing cholangitis syndrome (IHSC) syndrome
2674. ✓Gaucher syndrome type 2
2675. ✓Ichthyosis–hypotrichosis syndrome
2676. ✓Neonatal ichthyosis–sclerosing cholangitis (NISCH) syndrome
2677. ✓ keratitis–ichthyosis–deafness KID syndrome
2678. ✓ keratosis linearis with ichthyosis congenita and sclerosing keratoderma (klick) syndrome
2679. ✓Nephrosis with ichthyosis and adrenal insufficiency
2680. ✓Keratolysis exfoliativa congenita =continual peeling skin
2681. ✓Generalized inflammatory peeling skin syndrome= Peeling skin syndrome Type B
2682. ✓Generalized peeling skin syndrome= Peeling skin syndrome Type A
2683. ✓Acral peeling skin syndrome
2684. ✓Loricin Keratoderma
2685. ✓Infantile Gaucher disease
2686. ✓Refsum disease=heredopathis atactica polyneuritiformis
2687. ✓Sjögren-larsson syndrome

2688. ✓Dolichol kinase deficiency
2689. ✓ELOVL4 deficiency syndrome
2690. ✓Tay-sachs syndrome (IBIDS) (congenital ichthyosis with trichothiodystrophy) (Ichthyosis, Brittle hair, Intellectual impairment, Decreased fertility, and Short stature)
2691. ✓Progressive symmetric Erythrokeratoderma= Gottron syndrome= Darier -Gottron syndrome=Erythrokeratodermia figurata variabilis=Symmetrical progressive erythrokeratodermia=Progressive symmetric erythrokeratodermia
2692. ✓Erythrokeratoderma en cocardes
2693. ✓periorificial and ptychotropic erythrokeratoderma PERIOPTER syndrome
2694. ✓Reticular Erythrokeratoderma
2695. ✓Erythrokeratodermia with spinocerebellar ataxia (giroux–barbeau syndrome
2696. ✓Erythrokeratodermia-cardiomyopathy syndrome
2697. ✓Erythrokeratoderma variabilis EKV=mendes da costa disease=keratosis rubra figurate
2698. ✓Erythrokeratodermia variabilis Cram–Mevorah= Annular erythrokeratodermia figurata variabilis
2699. ✓ Erythrokeratodermia variabilis with erythema gyratum repens
2700. ✓KSDR-related erythrokeratoderma
2701. ✓ Acquired Ichthyosis
2702. Icterus marginatus
2703. Id reaction= autoeczematization=Autosensitization dermatitis
2704. Idiopathic dermal melanosis
2705. Idiopathic eruptive hypopigmentation
2706. Idiopathic eruptive macular pigmentation (IEMP)
2707. Idiopathic facial aseptic granuloma (IFAG)
2708. Idiopathic follicular hyperkeratotic spicules
2709. Idiopathic granulomatous mastitis (IGM)
2710. Idiopathic guttate hypomelanosis IGH=leukopathia symmetrica progressiva
2711. Idiopathic lobular panniculitis of pregnancy

2712. **Idiopathic lymphoplasmacellular mucositis-dermatitis**
2713. **Idiopathic mucositis**
2714. **Idiopathic orbital inflammatory syndrome**
2715. **idiopathic scrotal calcification**
2716. **Idiopathic thrombocytopenic purpura**
2717. **Idiopathic unilateral lower limb gangrene in a neonate**
2718. **Illusory pigmentation of pseudo-hutchinson's sign**
2719. **IL-10/IL-10r deficiency**
2720. **IL-1 receptor-associated kinase-4 (IRAK-4) deficiency**
2721. **Immersion foot= Trench foot**
2722. **Immunoglobulin g4-related disease (IGG4-RD)**
2723. **Immunodeficiency with lymphoid proliferation**
2724. **Immunoglobulin deficiency disorders (see next)**
2725. ✓ **Agammaglobulinemia**
2726. **X-linked agammaglobulinaemia (Bruton's disease)**
2727. ✓ **Common variable immunodeficiency**
2728. ✓ **Selective Ig A immunodeficiency**
2729. ✓ **Selective Ig M deficiency**
2730. ✓ **Tweak deficiency**
2731. ✓ **Activated pi3kδ syndrome**
2732. ✓ **x-linked hyper-igm syndrome=Class-switch recombination defects**
2733. ✓ **AR hyper-IgM syndromes**
2734. ✓ **hypohidrotic ectodermal dysplasia with immunodeficiency**
2735. ✓ **immunodeficiency, centromeric instability, and facial anomalies (ICF) syndrome**
2736. ✓ **Transient hypogammaglobulinemia of infancy**
2737. ✓ **Thymoma with immunodeficiency**
2738. **Impetigo (non-bullous & bullous)**
2739. **Impetigo circinate**
2740. **Impetigo of bochart=Superficial pustular folliculitis**
2741. **Impetigo herpetiformis**
2742. **Impetigo incognito**

2743. **Impetiginoid syphiloderm**
2744. **Impetiginization**
2745. **Incidentaloma**
2746. **Indian tick typhus**
2747. **Infantile digital fibromatosis = inclusion body fibromatosis**
2748. **Infantile gluteal granuloma (papuloerosive dermatitis of Jacquet and Sevestre)**
2749. **Infantile myofibromatosis**
2750. **Infantile pedal papules**
2751. **Infantile perineal (perianal) protrusion**
2752. **Infantile systemic hyalinosis**
2753. **Infantile transient smooth muscle contraction of the skin**
2754. **Infantile tremor syndrome**
2755. **Infective dermatitis: HTLV1**
2756. **Inflammatory pseudotumor**
2757. **Inflammatory papillary hyperplasia**
2758. **Incontinentia pigmenti=bloch–Sulzberger syndrome(inflammatory= vesicular & verrucous & hyperpigmented& hypopigmented=atrophic)**
2759. **Infundibuloma**
2760. **infundibular cyst**
2761. **Injection site reactions**
2762. **Interdigital blastomycetia erosio**
2763. **Interdigital candidiasis**
2764. **Interdigital erythrasma**
2765. **Interdigital gram negative toe web infection**
2766. **Interdigital intertrigo**
2767. **Interdigital pseudomonaus**
2768. **Interdigital tinea pedis**
2769. **Interferon-induced vitiligo**
2770. **Interferon-associated cutaneous necrosis**
2771. **Interstitial granulomatous dermatitis (ackerman syndrome)**
2772. **Intertrigo**

2773. **Intravascular papillary endothelial hyperplasia -masson's pseudoangiosarcoma**
2774. **Intravascular histiocytosis & intralymphatic histiocytosis (IH)**
2775. **Intraepithelial neoplasia**
2776. **Intrauterine epidermal necrosis**
2777. **Insect bite reaction**
2778. **Invasive fungal dermatitis**
2779. **Inverted follicular keratosis**

Inverse in dermatology (SEE NEXT)

2780. **✓Acne inversa**
2781. **✓Inverse gottron papules**
2782. **✓Inverse halo phenomenon**
2783. **✓Inverse klippel-trenaunay syndrome**
2784. **✓Inverse koebner phenomeno**
2785. **✓Inverse lichen planus**
2786. **✓Inverse lichenoid drug eruption**
2787. **✓Inverse papular acrokeratosis (acrokeratoelastoides of oswaldo costa)**
2788. **✓Inverse pityriasis rosea**
2789. **✓Inverse psoriasiform drug eruption**
2790. **✓Inverse psoriasis**
2791. **✓Inverse sporotrichoid pattern of cutaneous tuberculosis**
2792. **✓Inverse subcorneal pustular dermatosis**
2793. **✓Inverse tinea versicolor**
2794. **✓Inverse type of junctional epidermolysis bullosa (jeb inversa)**
2795. **✓Inverse type of recessive dystrophic epidermolysis bullosa (rdeb-i)**
2796. **✓Lichen planus pigmentosus (lpp) inversus**
2797. **✓Ophiasis inversus**
2798. **✓Pterygium inversum unguis (piu)**
2799. **Invisible dermatoses**
2800. **Iododerma**

2801. **IPEX syndrome (immune dysregulation, polyendocrinopathy, enteropathy, x-linked syndrome)**
2802. **Ipsilateral masseter's spasm**
2803. **Iron deficiency anemia related skin disorders**
2804. **Ischemia-induced perianal ulceration**
2805. **Ischemic fasciitis**
2806. **Isolated melanocytic hyperpigmentation of the distal digits**
2807. **Isolated occipital white lock**
2808. **Isolated polymyositis**
2809. **Isolated white forelock**
2810. **Isovaleric academia**
2811. **Israeli tick typhus**
2812. **Isthmicoma**
2813. **Isthmus-catagen cyst**
2814. **Itching purpura= disseminated pruriginous angiodermatitis**
2815. **Ito hypomelanosis=incontinentia pigment achromians=systematized achromic nevus**
2816. **Jackson-sertoli syndrome**
2817. **Jacquet erosive dermatitis**
2818. **Jaffe–campanacci syndrome**
2819. **Janeway lesions**
2820. **Japanese spotted fever=Oriental spotted fever**
2821. **Johanson–blizzard syndrome**
2822. **Johnson–mcmillin syndrome**
2823. **Jogger's nipples=Runner's nipples**
2824. **Joint contractures, muscle atrophy, microcytic anemia, and panniculitis-induced lipodystrophy (jmp) syndrome**
2825. **Juvenile aponeurotic fibroma**
2826. **Juvenile gangrenous vasculitis of the scrotum**
2827. **Juvenile hyaline fibromatosis**
2828. **Juvenile idiopathic arthritis**
2829. **Juvenile melanoma**
2830. **Juvenile temporal arteritis**

2831. **Juvenile plantar dermatosis=atopic winter feet=forefoot dermatitis =sweaty sock dermatitis=dermatitis plantaris sicca**
2832. **Juvenile polyposis syndrome**
2833. **Juvenile Spongiotic Gingivitis**
2834. **Juvenile spring eruption**
2835. **Juvenile xanthogranuloma**
2836. **Kabuki syndrome**
2837. **Kallin syndrome (epidermolysis bullosa simplex with anodontia or hypodontia, nail dystrophy and alopecia)**
2838. **Kanzaki syndrome**
2839. **Kaposi sarcoma** **(see next)**
2840. **✓Classic**
2841. **✓Bullous**
2842. **✓ Pyogenic granuloma-like kaposi sarcoma (pg-like ks)**
2843. **✓African endemic**
2844. **✓ Iatrogenic**
2845. **✓Immunosuppression related**
2846. **✓AIDS-related epidemic**
2847. **✓pseudo bullous lymphangioma like**
2848. **✓keliodal**
2849. **Kaposi dermatitis= Xeroderma pigmentosum**
2850. **Kaposiform hemangioendothelioma**
2851. **Kaposi-Irgang disease= lupus profundus**
2852. **Kaposi-Stemmer sign**
2853. **Kaposiform lymphangiomatosis**
2854. **Kaposi varicelliform eruption**
2855. **Kasabach merritt syndrome=hemangioma with thrombocytopenia**
2856. **Kawasaki disease =mucocutaneous lymph node syndrome= Kawasaki Syndrom**
2857. **Kayser-fleischer rings**
2858. **Kearns-sayre syndrome**
2859. **Keipert syndrome**

2860. **Keloid**
2861. **Ketron-goodman disease**
2862. **Keratin granuloma**
2863. **Keratoacanthoma= molluscum sebaceum (SEE next)**
2864. **✓Solitary**
2865. **✓Giant**
2866. **✓Nodular**
2867. **✓Subungual**
2868. **✓Mucosal**
2869. **✓Centrifugum marginatum**
2870. **✓ Generalized eruptive keratoacanthoma of grzybowski**
2871. **✓ Multiple keratoacanthomas ferguson-smith syndrome**
2872. **✓Chronic involuting relapsing keratoacanthoma**
2873. **✓Mixed syndrome =witten and zak syndrome**
2874. **Keratocyst= Isthmic-anagen cyst**
2875. **Keratoderma blennorrhagicum**
2876. **Keratoderma climatricum =Haxthausen's Disease**
2877. **Keratoderma simplex**
2878. **Keratolysis exfoliativa =dyshidrosis lamellosa sicca= recurrent focal palmar peeling**
2879. **Keratolysis exfoliativa congenita =continual peeling skin**
2880. **Keratolytic winter erythema=oudtshoorn skin=erythrokeratolysis hiemalis**
2881. **Keratosis alba**
2882. **Keratosis circumscripta**
2883. **Keratosis follicularis squamosa**
2884. **Keratosis lichenoid chronica =Nekam disease=Lichen verrucosus et reticularis=lichenoid trikeratosis=porokeratosis striata lichenoides=lichen ruber moniliformis= keratose lichenoide striae**
2885. **Keratosis multiformis**
2886. **Keratosis pilaris= Lichen pilaris**
2887. **Keratosis pilaris decalvans**
2888. **Keratosis pilaris rubra faceii**

2889. **Keratosis follicularis spinulosa decalvans KFSD**
2890. **Keratosis rubra pilaris faciei atrophicans**
2891. **Keratosis palmoplantaris nummularis=hereditary painful callosities**
2892. **Papular, profuse, precocious keratosis pilaris**
2893. **Keutel syndrome**
2894. **Killian syndrome**
2895. **Kikuchi disease**
2896. **Kimura's disease**
2897. **Kindler syndrome =Kindler–Weary syndrome= Bullous acrokeratotic poikiloderma of Kindler and Weary**
2898. **Kirghizian dermato-osteolysis**
2899. **Kirman syndrome**
2900. **Klinefelter syndrome**
2901. **Klippel–trénauna syndrome=(naevus vasculosus osteohypertrophicus; angioosteohypertrophy syndrome**
2902. **Klippel-feil syndrome**
2903. **Knuckle pads**
2904. **Kobberling-dunnigan syndrome**
2905. **Koebner phenomenon**
2906. **Koebner syndrome**
2907. **Koenen tumour**
2908. **Koro syndrome**
2909. **Krabbe's disease**
2910. **Kwashiorkor= Protein-calorie malnutrition**
2911. **Kyrle's disease= hyperkeratosis follicularis et parafollicularis in cutem penetrans**
2912. **Labrune syndrome**
2913. **L-2-hydroxyglutaric aciduria**
2914. **Lacrimo-auriculo-dento-digital syndrome**
2915. **Lafora's disease**
2916. **LAMB syndrome (Lentigines, Atrial myxomas, Mucocutaneous myxomas, Blue nevi)**
2917. **Late-onset focal dermal elastosis**

2918. **Lawrence syndrome**
2919. **Laddering melanotic pattern of langer's lines (LMPLL)**
2920. **Large cell acanthoma**
2921. **Large hematoma occlusion syndrome**
2922. **Larsen syndrome**
2923. **Larsen-like syndrome**
2924. **Larva currens**
2925. **Larva migran =cutaneous larvae migrans (CLM)=dermatitis verminosa serpiginosa=Creeping Eruption=plumber's itch**
2926. **Laser therapy– induced comedones**
2927. **Lassa fever**
2928. **Laugier-hunziker syndrome**
2929. **Launois-bensaude syndrome**
2930. **Leaflike denture fibroma**
2931. **Legius syndrome**
2932. **Leiomyoma cutis& Piloleiomyomas& Leiomyomas of the Genital Skin**
2933. **Leiomyosarcoma**
2934. **Leigh syndrome**
2935. **Leiner's disease= erythroderma desquamativum**
2936. **Leishmaniasis (see next)**
2937. **Old World cutaneous leishmaniasis =Baghdad boil= oriental sore (see next)**
2938. **Zoonotic**
2939. **Anthroponotic**
2940. **Leishmaniasis Recidivans=Chronic Lupoid Leishmaniasis**
2941. **Lupoid**
2942. **Dermatomal (Zosteriform) Cutaneous Leishmaniasis**
2943. **Erysipelas - Like Cutaneous Leishmaniasis (Erysipeloid Type)**
2944. **Verrucous Cutaneous Leishmaniasis (Warty CL)**
2945. **Hyperkeratotic Like Cutaneous Leishmaniasis**
2946. **Tumor-Like Cutaneous Leishmaniasis(Pseudo-Tumoral CL)**

2947. **New World cutaneous leishmaniasis= American Leishmaniasis**
2948. ✓ **Sarcoid-like**
2949. ✓**lupoid**
2950. ✓**Ulcerated**
2951. ✓**Vegetating**
2952. ✓**Erysipeloid**
2953. ✓**Sporotrichoid**
2954. ✓**Impetigo-like**
2955. ✓**verrucous**
2956. ✓**pustular**
2957. ✓**plaque-like**
2958. ✓**Eczematoid**
2959. ✓**Furunculoid**
2960. ✓**Miliary**
2961. ✓**Lichenoid**
2962. ✓**papulotuberous**
2963. ✓**Leishmanid**
2964. ✓**Mucocutaneous**
2965. ✓**Mucosal leishmaniasis**
2966. ✓**Disseminated**
2967. ✓**Diffuse Cutaneous Leishmaniasis= anergic and lepromatous leishmaniasis = pseudolepromatous leishmaniasis= Lepromatoid Leishmaniasis**
2968. ✓**Visceral= kala-azar=dum-dum fever**
2969. ✓**post kala-azar Dermal leishmaniasis (PKDL)**
2970. **Lenaerts syndrome**
2971. **Inherited pattern Lentigenossis**
2972. **Acral lentigines (AL)**
2973. **Centrofacial Lentiginosis**
2974. **Eruptive lentiginosis**
2975. **Generalized Lentiginosis**

2976. **Lentiginosis : oral melanotic macules & labial melanotic macules & genital lentiginosis**
2977. **Lentiginous dysplastic nevus of elderly**
2978. **Puva- induced lentigines**
2979. **Segmental lentigenosis= unilateral lentigenosis= partial zosteriform lentiginosis**
2980. **Lentigo simplex**
2981. **Solar Lentigo =senile freckle**
2982. **Ink spot Lentigo**
2983. **Lenz-Majewski syndrome**
2984. **Leopard syndrome (Lentigines , ECG abnormalities, Ocular hypertelorism , Pulmonary valve stenosis , Abnormal genitalia, Retardation of growth , and Deafness)**
2985. **Leprechaunism (donohue)**
2986. **Leprosy =Hansen disease=Hanseniasis=Elephantiasis Graecorum (see next)**
2987. **✓ Borderline leprosy (BT&BL&BB): Borderline Tubercloid & Borderline Lepromatous &Mid borderline**
2988. **✓Dimorphous leprosy**
2989. **✓ Histoid leprosy**
2990. **✓Lazarine leprosy**
2991. **✓ Indeterminate leprosy**
2992. **✓ Lepromatous leprosy LL**
2993. **✓Pure Neuritic Leprosy**
2994. **✓ Tubercloid leprosy TT**
2995. **✓Leprosy incognito**
2996. **✓Lucio leprosy=diffuse lepromatous leprosy =Pretty leprosy=lepra bonita**
2997. **✓ Lucio phenomenon**
2998. **✓Exacerbation reaction**
2999. **✓Type 1 reversal reaction T1R**
3000. **✓Type 2 leprosy reaction= erythema nodosum leprosum T2R**

3001. ✓Erythema nodosum leprosum(necrotican &ulcerative& bullous&Pustular&Target like)
3002. ✓ Verrucous leprosy=verrucous leproma
3003. ✓Leprosy deformity /deformities/disabilities
3004. ✓Rhinomaxillary syndrome of leprosy
3005. Leptospirosis
3006. Leucinosis
3007. Leucocyte adhesion deficiency – LAD (3 types)
3008. Leukaemia cutis
3009. Leukocytoclastic vasculitis=cutaneous small vessel vasculitis
3010. Leukodema
3011. Leukoderma due to drug
3012. Chemical leukoderma
3013. Contact leukoderma (contact vitiligo, chemical leukoderma& chemical or contact depigmentation)
3014. leukoderma punctata
3015. Leukoderma syphiliticum
3016. Leukoencephalopathy, brain calcifications, and cysts
3017. Leukoplakia
3018. Proliferative Verrucous Leukoplakia
3019. Oral hairy leukoplakia
3020. Leukoplakia with tylosis and esophageal carcinoma
3021. Leukotrichia
3022. Levamisol =cocaine vasculopathy/levamisole-induced vasculopathy/vasculitis
3023. Lichen amyloidosis
3024. Lichen aureus= lichen purpuricus
3025. Lichen dermatitis
3026. Lichen follicularis tumidus
3027. Lichen nitidus(generalized & perforating &nail &spinous)
3028. Actinic lichen nitidus = summertime actinic lichenoid eruption
3029. Lichen nuchae

3030. **Lichen pilaris**
3031. **Lichen planus (see next) LP**
3032. **✓ Atrophic**
3033. **✓ Annular**
3034. **✓ Acute exanthematous**
3035. **✓ Hypertrophic**
3036. **✓ Bullous Lichen planus**
3037. **✓ Lichen planus pemphigoides**
3038. **✓ Actinic = Lichen planus actinicus= lichenoid melanodermatitis= LP tropicus**
3039. **✓ Inverse**
3040. **✓ Lichen planus pigmentosus**
3041. **✓ Linear**
3042. **✓ Zosteriform**
3043. **✓vulvovaginal gingival syndrome**
3044. **✓Guttate**
3045. **✓ Follicular**
3046. **✓Ulcerative LP**
3047. **✓ Lichen planus –DLE overlap**
3048. **✓oral Lichen planus OLP (ulcerative & erosive &reticulate &atrophy &papule &plaque&bullous&pigmented& Mixed)**
3049. **✓vulvovaginal lp**
3050. **✓Lichen planus poikiloderma**
3051. **Lichen planopilaris**
3052. **Lichen Planopilaris Pemphigoides**
3053. **Lichen ruber acuminatus**
3054. **Lichen ruber moniliformis**
3055. **✓Lichen sclerossus et atrophicus LSEA= Lichen albus**
3056. **✓Lichen sclerossus extra genital**
3057. **✓Lichen sclerossus bullous hemorrhagic**
3058. **✓Hyperkeratotic lichen sclerossus**

3059. ✓Lichen sclerossus et atrophicus /morphea overlap
3060. Lichen simplex chronicus LSC
3061. Giant lichenification of Pautrier
3062. Lichen spinulosus
3063. Lichen urticatus
3064. Lichen verrucosus et reticularis
3065. Lichenoid and granulomatous stomatitis
3066. Oral Lichenoid Contact Lesions
3067. Oral Lichenoid Drug Reactions
3068. Oral lichenoid lesions Related to Graft-Versus- Host Disease (OLL-GvHD)
3069. Lichenoid keratosis=lichen planus-like keratoses
3070. Lichenoid granulomatous stomatitis
3071. Lichenoid granulomatous dermatitis
3072. Lichenoid Dysplasia (LD)
3073. Lichenoid pseudovesicular papular eruption
3074. Ligature injury
3075. Limb girdle muscular dystrophy
3076. Limb–mammary syndrome
3077. Linea alba
3078. Linear ectodermal cutaneous hamartoma
3079. Linear IG A bullous dermatosis LABD= IG A bullous pemphigoid=linear dermatitis herpetiformis=linear IG A dermatosis (lad)
3080. Linear naevus syndrome
3081. Linea nigra
3082. Linea fusca (lf)
3083. Linear gingival erythema
3084. Linear nevoid hypopigmentation
3085. Linear and whorled nevoid hypermelanosis= linear nevoid hyperpigmentation = zebra-like hyperpigmentation in whorls and streaks
3086. Linear focal elastosis

Lines in dermaology (see next)

3087. ✓Antecubital ahn's line

3088. ✓Aldrich-Mees' lines
3089. ✓Beau's lines
3090. ✓Bismuth line
3091. ✓Bunny lines
3092. ✓Blaschko's lines
3093. ✓Burton line=lead line
3094. ✓Dennie–morgan lines/folds
3095. ✓Drug line
3096. ✓Embryonic milk line
3097. ✓Fingerprint lines
3098. ✓Glabellar frown lines
3099. ✓Hart's line
3100. ✓Horizontal necklace lines
3101. ✓Juxtaclavicular beaded lines
3102. ✓kraissl's lines
3103. ✓Marionette lines
3104. ✓Mees' lines
3105. ✓Muehrcke's lines
3106. ✓pastia's lines
3107. ✓pigmentary demarcation lines
3108. ✓Relaxed skin tension lines
3109. ✓Sleep lines
3110. ✓Transverse nasal line
3111. ✓Wallace line
3112. ✓Yellow line of pinkus
3113. ✓Hinderer's lines
3114. Lingual papillitis
3115. Lingual thyroid
3116. Lingual tonsil hypertrophy
3117. Lingual epidermal choristoma
3118. Lip lick dermatitis

3119. **Lip pit**
3120. **Lipedema**
3121. **Lipodermatosclerosis =hypodermitis sclerodermaformis= sclerosing panniculitis=stasis panniculitis**
3122. **Lipodystrophy**
3123. **✓lipoatrophia semicircularis**
3124. **✓Annular lipoatrophy**
3125. **✓Congenital generalized lipodystrophy**
3126. **✓berardinelli-seip congenital lipodystrophy (BSCL)**
3127. **✓familial partial lipodystrophy(dunnigan variant&kobberling variant)**
3128. **✓Familial partial lipodystrophy associated with mandibuloacral dysplasia**
3129. **✓Acquired generalized lipodystrophy**
3130. **✓Acquired partial lipodystrophy barraquer–simons syndrome**
3131. **✓lipodystrophia centrifugalis abdominalis infantilis**
3132. **✓HIV/ART-associated lipodystrophy syndrome**
3133. **✓Recurrent lipoatrophic panniculitis of children**
3134. **✓lipoatrophia annularis (ferreira-marques syndrome)**
3135. **✓localized lipoatrophy**
3136. **✓Localized involutional lipoatrophy**
3137. **✓lipohypertrophy**
3138. **✓Insulin lipodystrophy**
3139. **✓Hemi lipodystrophy**
3140. **Levator ani syndrome**
3141. **Lhermitte-duclos disease variant of pten hamartoma syndrome**
3142. **Lipedematous alopecia**
3143. **Lipedematous Scalp**
3144. **Lipogranulomatosis of penis**
3145. **Lipoid proteinosis= hyalinosis cutis et mucosae=Urbach – Wiethe syndrome**
3146. **Lipoma**
3147. **Lipomatosis**
3148. **Lipomatosis of nerve**

3149. **Lipoblastoma**
3150. **Lipoblastomatosis**
3151. **Lipoblastoma-like tumor of the vulva**
3152. **Lipofibromatosis**
3153. **Lipofibromatosis-like neural tumor**
3154. **Lipogranulomatosis**
3155. **Liponeurofibroma**
3156. **Liposarcoma**
3157. **Spindle cell liposarcoma**
3158. **Myxoid liposarcoma**
3159. **Lichtenberg figures**
3160. **Listeriosis**
3161. **Livedo reticularis**
3162. **Livedo racemosa**
3163. **Livedoid vasculopathy = atrophie blanche**
3164. **Liver diseases &hepatitis &cirrhosis skin signs**
3165. **Lobomycosis= Lobo's disease= keloidal blastomycosis=Lacaziosis**
3166. **Localized hypopigmented keratoses**
3167. **Locus minoris resistentiae**
3168. **Loeffler syndrome**
3169. **Loaiasis= Loa loa filariasis**
3170. **Loeys–dietz syndrome**
3171. **Löfgren syndrome**
3172. **Loose anagen syndrome**
3173. **Loxoscelism**
3174. **Lubac (linear ubiquitination assembly complex) components deficiency**
3175. **Lucky winter syndrome**
3176. **Ludwig angina**
3177. **Lues maligna of syphilis**
3178. **Lupus erythematosus (see next)**
3179. **✓Acute cutaneous lupus erythematosus (ACLE) :localized or generalized erythema or bullae,generally associated with SLE**

3180. ✓Subacute lupus (papulosquamous= psoriasiform & annular =lupus marginatus=lupus erythematosus gyratum repens)
3181. ✓Drug-induced lupus erythematosus
3182. ✓Acute syndrome of apoptotic pan-epidermolysis= Toxic epidermal necrolysis-like acute cutaneous lupus erythematosus
3183. ✓ Bullous lupus erythematous
3184. ✓ chilblain lupus
3185. ✓ DLE discoid lupus erythematosus (localized& disseminated)
3186. ✓Comedogenic lupus
3187. ✓Pigmented type of chronic cutaneous lupus erythematosus
3188. ✓ verrucous discoid lupus erythematosus
3189. ✓ Hypertrophic lupus
3190. ✓Lichenoid discoid lupus erythematosus
3191. ✓ Neonatal lupus erythematous
3192. ✓lupus panniculitis
3193. ✓lupus profundus
3194. ✓Mucosal discoid lupus erythematosus
3195. ✓lupus erythematosus tumidus
3196. ✓ Systemic lupus erythematous SLE
3197. Lupus miliaris disseminatus facei. LMDF
3198. Lyell's syndrome =toxic epidermal necrolysis
3199. Lyme disease
3200. Lymphangitis
3201. lymphangitis carcinomatosis
3202. Nodular lymphangitis syndrome
3203. Lymphangiosarcoma= Angiosarcoma
3204. Lymphangioma
3205. Lymphangioma circumscriptum=microcystic lymphatic malformation
3206. Macrocystic lymphatic malformation=cystic hygroma= lymphangioma cysticum
3207. Combined microcystic and macrocystic lymphatic malformation
3208. Lymphangiomatosis

3209. **Lymphangiomyomatosis**
3210. **Kaposiform lymphangiomatosis**
3211. **Lymphangioleiomyomatosis**
3212. **Lymphangiopericytoma**
3213. **Lymphangiectasia**
3214. **Benign Lymphangioendothelioma=Acquired progressive lymphangioma**
3215. **Net-like superficial lymphatic malformation**
3216. **Lymphedema**
3217. **Congenital onset of primary lymphoedema=Milroy disease**
3218. **Primary lymphoedema with later onset=Meige disease**
3219. **Milroy-like disease**
3220. **Hereditary lymphedema**
3221. **Lymphedema-Atrial Septal Defects-Facial Changes syndrome=Irons-Bianchi syndrome**
3222. **Lymphoedema – myelodysplasia =emberger syndrome**
3223. **Microcephaly with or without chorioretinopathy lymphedema or impaired intellectual development**
3224. **Hennekam lymphangiectasia lymphedema syndrome**
3225. **lymphedema-choanal atresia**
3226. **Hypotrichosis–lymphedema–telangiectasia syndrome**
3227. **Lymphoedema–lipoedema syndrome**
3228. **Massive localized lymphedema**
3229. **Lymphedema praecox**
3230. **Lymphedema tarda**
3231. **Factitial lymphedema**
3232. **Lymphedema–distichiasis syndrome**
3233. **Lymphedema: filariasis=Elephantiasis tropica (elephantiasis arabum)**
3234. **Nodular lymphedema**
3235. **Cholestasis – lymphoedema syndrome (aagenaes syndrome)**
3236. **Generalized, multisegmental lymphatic dysplasia**
3237. **osteopetrosis, lymphoedema, ectodermal dysplasia anhydrotic and immune deficiency syndrome(OLEDAID syndrome)**
3238. **Generalized lymphatic dysplasia (hennekam syndrome)**

3239. **Microcephaly – lymphoedema – chorioretinal dysplasia**
3240. **Primary lymphedema associated with yellow nails and pleural effusion(yellow nail syndrome)**
3241. **Lymphocele**
3242. **Lymphogranuloma venereum= Lymphogranuloma inguinale**
3243. **Lymphoepithelioma-like carcinoma**
3244. **Adult T-cell leukemia/lymphoma**
3245. **Follicular helper T-cell (TFH) lymphoma**
3246. **Angiocentric T-cell lymphoma**
3247. **Angioimmunoblastic T-cell lymphoma= Angioimmunoblastic lymphadenopathy= Lymphogranulomatosis X**
3248. **CD30-Negative Cutaneous T-Cell Lymphomas**
3249. **CD8-positive pleomorphic small/medium-sized T-cell lymphoma**
3250. **Hydroa vacciniforme–like lymphoma**
3251. **Juvenile granulomatous cutaneous lymphoma (JGCTL)**
3252. **Lymphomatoid papulosis (LYP)**
3253. **Pleomorphic T-cell lymphoma**
3254. **Primary cutaneous acral CD8-positive T-cell lymphoma PCATCL**
3255. **Primary cutaneous anaplastic large cell lymphoma (PC-ALCL)**
3256. **Primary cutaneous CD30-positive lymphoproliferative disorders (LYP)+(PC-ALCL)**
3257. **Primary cutaneous CD4-positive small/medium T-cell lymphoproliferative disorder PCSMTCLPD**
3258. **Primary cutaneous CD8+ aggressive epidermotropic cytotoxic T-cell lymphoma PCAETCL**
3259. **Primary cutaneous gamma/delta γ/δ T-Cell t-cell lymphoma**
3260. **Primary cutaneous peripheral T-cell lymphoma , unspecified**
3261. **Subcutaneous panniculitis-like T-cell lymphoma (SPTCL)**
3262. **Lennert's lymphoma (lymphoepithelioid lymphoma)**
3263. **Lymphoma T cell other ,NOS**
3264. **T cell prolymphocytic leukemia**
3265. **Intravascular T- and NK-cell lymphoma**

3266. **Mycosis fungoides MF (SEE NEXT)**
3267. ✓ **Hypopigmented Mycosis fungoides**
3268. ✓ **Pigmented purpura –like Mycosis fungoides**
3269. ✓ **Acanthosis nigricans like Mycosis fungoides**
3270. ✓ **Anetodermic Mycosis fungoides**
3271. ✓ **Bullous Mycosis fungoides**
3272. ✓ **Dyshidrosiform mycosis fungoides**
3273. ✓ **Erythema annulare centrifugum like mycosis fungoides**
3274. ✓ **Erythema gyratum repens-like mycosis fungoides (EGR MF)**
3275. ✓ **Folliculotropic mycosis fungoides/ pilotropic**
3276. ✓ **Granuloma annulare like Mycosis fungoides**
3277. ✓ **Granulomatous Mycosis fungoides**
3278. ✓ **Granulomatous slack skin**
3279. ✓ **Hyperpigmented Mycosis fungoides**
3280. ✓ **Ichthyosiform Mycosis fungoides**
3281. ✓ **Interstitial Mycosis fungoides**
3282. ✓ **Invisible mycosis fungoides**
3283. ✓ **keratosis lichenoides like Mycosis fungoides**
3284. ✓ **Poikiloderma vasculare atrophicans=Poikilodermatous mf**
3285. ✓ **Mycosis fungoides associated follicular mucinosis**
3286. ✓ **Mycosis fungoides palmaris et plantaris**
3287. ✓ **Papillomatous/ vegetating**
3288. ✓ **papular mycosis fungoides**
3289. ✓ **perioral dermatitis like mycosis fungoides**
3290. ✓ **pityriasis lichenoides like mycosis fungoides**
3291. ✓ **Psoriasiform mycosis fungoides**
3292. ✓ **pustular Mycosis fungoides**
3293. ✓ **pyoderma gangrenosum like – Mycosis fungoides**
3294. ✓ **Syringotropic mycosis fungoides**
3295. ✓ **Adnexotropic (folliculotropic and syringotropic)**
3296. ✓ **Unilesional/Solitary Mycosis fungoides**

3297. ✓ Verrucous/ hyperkeratotic Mycosis fungoides
3298. ✓Erythema chronicum migrans like- t cell lymphoma
3299. ✓Zosteriform mycosis fungoides
3300. ✓ pagetoid reticulosis (ketron-goodman disease& Woringer -kolopp)
3301. Sezary syndrome
3302. Extranodal natural killer (NK)/ T-cell lymphoma (ENKL)
3303. Blastic NK-cell lymphoma=blastic plasmacytoid dendritic cell neoplasm BPDCN= CD4+／CD56+ haematodermic neoplasm
3304. Aggressive NK-cell leukemia (ANKL)
3305. Intravascular T- and NK-cell lymphoma
3306. **B cell Lymphoma (see next)**
3307. ✓ Intravascular diffuse large B-cell lymphoma IVLBCL
3308. ✓ Primary cutaneous diffuse large B cell lymphoma, leg type PCDLBCL
3309. ✓Primary cutaneous follicle centre lymphoma PCFCL
3310. ✓ Cutaneous extranodal marginal zone lymphoma of mucosal-associated lymphoid tissue (MALT)
3311. ✓Primary cutaneous marginal zone lymphoma /lymphoproliferative disorder PCMZLPD
3312. ✓primary cutaneous marginal zone lymphoma (PCMZL)
3313. ✓Secondary cutaneous marginal zone lymphoma (scMZL)
3314. ✓Cutaneous marginal zone lymphomas CMZLs
3315. ✓Anetodermic form of primary cutaneous marginal zone lymphoma
3316. ✓ Primary cutaneous diffuse large B-cell lymphoma, other
3317. ✓B-chronic lymphocytic leukemia/small lymphocytic lymphoma CLL/SLL
3318. EBV-positive mucocutaneous ulcer EBVMCU
3319. EBV-associated diffuse large B-cell lymphoma of the elderly
3320. EBV-positive T- and NK-cell lymphoid proliferations and lymphomas of childhood
3321. EBV+ diffuse large B-cell lymphoma,not otherwise specified
3322. Primary cutaneous mantle cell lymphoma
3323. Precursor T Lymphoblastic leukemia/lymphoma
3324. Precursor B lymphoblastic leukemia/lymphoma

3325. **NK-lymphoblastic leukemia/lymphoma**
3326. **Cutaneous T Lymphoblastic lymphoma**
3327. **Cutaneous B Lymphoblastic lymphoma**
3328. **Lymphomatoid granulomatosis LG**
3329. **Cutaneous amyloidoma**
3330. **Composite lymphomas**
3331. **T-cell/histiocyte-rich B-cell lymphoma**
3332. **Primary effusion lymphoma**
3333. **Cutaneous lymphoid hyperplasia =pseudolymphoma= lymphocytoma cutis= spiegler–fendt sarcoid**
3334. **Predominant T-cell pseudolymphoma**
3335. **Predominant B-cell pseudolymphoma**
3336. **Borrelial lymphocytoma cutis = lymphadenosis benigna cutis**
3337. **Acral pseudolymphomatous angiokeratoma APACHE**
3338. **Pseudolymphoma: miliarial type**
3339. **T-cell rich angiomatoid polypoid pseudolymphoma**
3340. **Drug induced pseudolymphoma**
3341. **Lymphocytic Infiltrate of Jessner= Benign Lymphocytic Infiltrate of the Skin**
3342. **Immunocytoma**
3343. **Extramedullary hematopoiesis**
3344. **Waldenström's macroglobulinemia/ lymphoplasmacytic lymphoma**
3345. **Pyogenic lymphoma (neutrophil-rich cd30 anaplastic large cell lymphoma)**
3346. **Autoimmune lymphoproliferative syndrome (Canale–Smith syndrome)**
3347. **EBV-associated lymphoproliferative syndrome**
3348. **Post-transplant lymphoproliferative disorders**
3349. **Methotrexate-associated lymphoproliferative disorder**
3350. **Nodular hyperplasia**
3351. **Lymphoma en cuirasse**
3352. **Plasmacytoma**
3353. **Plasmablastic lymphoma**
3354. **Burkitt lymphoma**
3355. **Hodgkin lymphoma**
3356. **Acute Lymphocytic Leukemia ALL**

3357. **Acute Myeloid Leukemia AML**
3358. **Chronic Lymphocytic Leukemia CLL**
3359. **Chronic Myeloid Leukemia CML**
3360. **T-cell prolymphocytic leukemia (T-PLL)**
3361. **B-cell prolymphocytic leukaemia**
3362. **Hairy cell leukemia**
3363. **pretibial lymphoplasmacytic plaque**
3364. **Lymphoepithelial cyst**
3365. **Lymphoplasmacytic plaque in children(LPC)**
3366. **Lymphorrhoea**
3367. **Lesch -Nyhan syndrome**
3368. **Lynch syndrome**
3369. **Macrocephaly–cutis marmorata telangiectatica congenita**
3370. **MACS (macrocephaly, alopecia, cutis laxa, and scoliosis)**
3371. **Macrodystrophia lipomatosa**
3372. **Macroglossia**
3373. **Macrophage activation syndrome**
3374. **Macular lymphocytic arteritis=lymphocytic thrombophilic arteritis**
3375. **Maculae cerulae**
3376. **Madelung diseases=Benign symmetric lipomatosis**
3377. **MAGIC syndrome (mouth and genital ulcers with inflammed cartilage)**
3378. **Majeed syndrome**
3379. **Majocchi granuloma=Nodular granulomatous perifolliculitis**
3380. **Maffucci syndrome**
3381. **Malakoplakia = Malacoplakia**
3382. **Malaria**
3383. **Malignant endovascular papillary angioendothelioma =Dabska tumour**
3384. **Malignant endothelial papillary angioendothelioma**
3385. **Malignant fibrous histiocytoma**
3386. **Malignant granular cell tumor**
3387. **Malignant mixed tumor= malignant chondroid syringoma**
3388. **Mabry syndrome**
3389. **Mammary–digital–nail syndrome**

3390. **Mammary-type myofibroblastoma of soft tissue**
3391. **Mammary-type secretory carcinoma of the skin**
3392. **Mandibuloacral macrocephaly, alopecia, cutis laxa and scoliosis (MAC)**
3393. **Mandibuloacral dysplasia**
3394. **Mandibulofacial dysostosis**
3395. **Mannosidosis**
3396. **B-mannosidase deficiency**
3397. **Mantleoma**
3398. **Maple syrup urine disease**
3399. **Marantic endocarditis**
3400. **Marasmus**
3401. **Marfan syndrome**
3402. **Marfanoid progeria–lipodystrophy syndrome**
3403. **Marfanoid hypermobility syndrome**
3404. **Martinez syndrome**
3405. **Marinesco-sjogren syndrome**
3406. **Marshal –smith syndrome**
3407. **Marshal white syndrome**
3408. **Mastocytosis (see next)**
3409. ✓ **Adult type mastocytosis**
3410. ✓ **Telangiectasia macularis eruptive perstan TMEP**
3411. ✓ **Mastocytoma**
3412. ✓ **Nodular type**
3413. ✓ **Telangectatic**
3414. ✓ **Diffuse cutaneous mastocytosis**
3415. ✓ **Bullous type**
3416. ✓ **Xanthelasmoid= pseudoxanthomatous**
3417. ✓ **Urticaria pigmentosa=Maculopapular CM**
3418. ✓ **Mast cell leukemia**
3419. ✓ **Mast cell sarcoma**
3420. ✓ **Systemic mastocytosis (Indolent&Aggressive)**

3421. ✓ Systemic mastocytosis with associated clonal hematological non-mast cell-lineage disease (sm-ahnmd)
3422. Materia alba
3423. Maturational hyperpigmentation
3424. MAUIE syndrome (micropinnae, alopecia universalis, ichthyosis, and ectropion)
3425. McCune-albright syndrome
3426. Mcgrath syndrome= skin fragility syndrome
3427. Mckusick syndrome
3428. MDPL :mandibular hypoplasia, deafness, progeroid features, and lipodystrophy
3429. Measles=Rubeola= First Disease
3430. Mechanic's hands
3431. Mechanical trauma-induced comedones
3432. Medalion like dendrocyte hamartoma=CD34+ plaque-like dermal fibroma
3433. Median lip fissure
3434. Median raphe cyst
3435. Mediterranean Spotted Fever
3436. Mednik syndrome(mental retardation, enteropathy, deafness, peripheral neuropathy, ichthyosis, keratodermia)
3437. Megalencephaly capillary malformation MCAP
3438. Megalencephaly-polydactyly-polymicrogyria-hydrocephalus (MPPH) syndrome
3439. Megarbane–loiselet neonatal progeroid
3440. Meige disease
3441. Melnick–Fraser syndrome
3442. Melkersson–rosenthal syndrome
3443. Mendes da costa syndrome
3444. Melanocytic matricoma
3445. Melanosis of the nipple and areola
3446. Melanosis Universalis Hereditaria
3447. Melanoleucoderma
3448. Melanoacanthoma

3449. **Melanoma (see next)**
3450. ✓ **Acral lentiginous melanoma**
3451. ✓ **Amelanotic melanomas**
3452. ✓ **Balloon cell melanoma**
3453. ✓ **Borderline melanoma& Nevoid melanoma**
3454. ✓ **Malignant blue nevus =blue nevus like melanoma**
3455. ✓ **Clear cell sarcoma=Malignant Melanoma of Soft Tissue**
3456. ✓ **Desmoplastic melanoma**
3457. ✓ **Lentigo maligna**
3458. ✓ **lentigo maligna melanoma**
3459. ✓ **Melanoma associated hyperpigmentation MAH**
3460. ✓ **Melanoma induced leukoderma**
3461. ✓ **Melanoma metastasis**
3462. ✓ **Blue nevus-like metastatic melanoma**
3463. ✓ **Mucosal melanoma**
3464. ✓ **Nested melanoma of the elderly (NME)**
3465. ✓ **Nodular melanoma**
3466. ✓ **Polypoid nodular melanoma**
3467. ✓ **Pigmented epithelioid melanocytomas=Animal type melanoma**
3468. ✓ **Primary dermal melanoma**
3469. ✓ **Small cell melanoma**
3470. ✓ **Spitzoid melanoma**
3471. ✓ **Subungual melanoma**
3472. ✓ **Superficial spreading melanoma**
3473. ✓ **Melanocarcinoma**
3474. ✓ **childhood melanoma**
3475. ✓ **Melanoma in situ**
3476. ✓ **Recurrent melanoma**
3477. ✓ **Verrucous melanoma**
3478. ✓ **MELTUMP (melanocytic tumors of uncertain malignant potential)**

3479. ✓Superficial atypical melanocytic proliferation of uncertain significance (SAMPUS)
3480. ✓Intraepidermal atypical melanocytic proliferation of uncertain significance (IAMPUS)
3481. Melanotic neuroectodermal tumor
3482. MELAS: mitochondrial encephalomyopathy, lactic acidosis and stroke-like episodes
3483. Melasma =chloasma=Mask of pregnancy
3484. Extra-facial melasma
3485. 308-nm uvb induced melasma
3486. Melioidosis
3487. Melorheostosis
3488. MEND syndrome (male EBP disorder with neurologic defects)
3489. Meningococcemia
3490. Meningomyeloceles
3491. Meningocele
3492. Menkes syndrome
3493. Mental retardation, enteropathy, deafness, neuropathy, ichthyosis, keratodermia
3494. Meralgia paresthetica
3495. Merkel cell carcinoma =cutaneous apudoma= primary neuroendocrine carcinoma of the skin
3496. Mesonephric cyst
3497. Mesothelial cyst
3498. Mesotherapy related complications
3499. Metageria
3500. Metastasis & metastases
3501. Methylmalonic acidemia
3502. Methylmalonic aciduria
3503. 3-methylcrotonylcoa carboxylase (ec 6.4.1.4) deficiency
3504. Metophyma
3505. Mevalonate kinase deficiency
3506. Mikulicz syndrome

3507. **Microcephaly-capillary malformation syndrome**
3508. **Microcephaly-intracranial calcification syndrome**
3509. **Michelin tire baby =Folded skin with scarring =circumferential symmetric skin creases**
3510. **Microcystic adnexal carcinoma**
3511. **Microscopic polyangitis= microscopic polyarteritis**
3512. **Microglossia**
3513. **Microvenular hemangioma**
3514. **MIDAS : (Microphthalmia, Dermal Aplasia, and Sclerocornea) =Microphthalmia with linear skin defects(MLS) syndrome**
3515. **Midface toddler excoriation syndrome MiTES**
3516. **Midline anterior neck inclusion cyst**
3517. **Midline skin tag**
3518. **Midline upper lip sinus**
3519. **Mid dermal elastolysis**
3520. **Miescher's radial granulomas**
3521. **Migratory thrombophlebitis**
3522. **Milia & Milium Cysts**
3523. **Milia en plaque**
3524. **Milia like calcinosis**
3525. **Transverse nasal crease milia**
3526. **Milian citrine skin**
3527. **Miliaria=Heat rash=prickly heat**
3528. **Miliaria crystallina**
3529. **Miliaria rubra**
3530. **Miliaria profunda**
3531. **Miliaria pustulosa**
3532. **Miliaria scarlatinosa**
3533. **Miliary haemangiomatosis of infancy**
3534. **Miliary gout**
3535. **Miliary osteomata cutis**
3536. **Miliary tuberculosis**
3537. **Milker nodule= paravaccinia=Pseudocowpox**

3538. **Millipedes**
3539. **Milroy's disease**
3540. **Mitochondrial cytopathies**
3541. **Mitochondrial respiratory chain disorder**
3542. **Mitten-line hyperpigmentation**
3543. **Mixed connective tissue disease(sharp syndrome)**
3544. **Mixed immunobullous disease of childhood**
3545. **Molluscum contagiosum**
3546. **Gianotti-Crosti syndrome-like reaction to molluscum**
3547. **Molluscum-induced comedone**
3548. **Molluscum leprosum**
3549. **Molluscum like histiocytosis**
3550. **Molluscoid Pseudotumor**
3551. **Molluscum fibrosum gravidarum**
3552. **Molluscum pendulum**
3553. **Molluscum sebaceum**
3554. **Mondor's disease= Sclerosing lymphangitis of the penis**
3555. **Mongolian spot =Dermal melanocytosis**
3556. **Monilthrix**
3557. **Monkeypox- Mpox**
3558. **Monogenic nod-like receptor family pyrin domain containing 1 (nlrp1)-associated diseases**
3559. **Montgomery tubercle**
3560. **Montgomery syndrome**
3561. **Moraxella infection**
3562. **Morbihan's disease=solid facial edema**
3563. **MORFAN syndrome (M:mental retardation & O:over growth & RF:remarkable facies & AN:acanthosis negrican)**
3564. **Morgagni-stewart-morel syndrome**
3565. **Morgellon disease**

3566. **Morphea =localized scleroderma (see next)**
3567. **✓Plaque-form**
3568. **✓Superficial**
3569. **✓Guttate**
3570. **✓ Deep=profunda**
3571. **✓keloidal**
3572. **✓Bullous**
3573. **✓ Linear**
3574. **✓En coup de sabre**
3575. **✓parry-romberg (hemifacial atrophy)**
3576. **✓Generalized**
3577. **✓ Disabling pansclerotic morphea of children**
3578. **✓ Postirradiation morphea**
3579. **Morsicatio buccarum**
3580. **Morsicatio linguarum**
3581. **Morel-lavallee lesion**
3582. **morton neuroma**
3583. **Morvan syndrome**
3584. **Mosaic acral keratosis**
3585. **Mosaic trisomy 8**
3586. **Mosaicism**
3587. **Moynahan syndrome= Xeroderma–talipes–enamel defect**
3588. **Mucinosis (see next)**
3589. **✓Scleromyxoedema =papular mucinosis**
3590. **✓Cutaneous myxoma**
3591. **✓ Cutaneous Focal mucinosis**
3592. **✓Follicular mucinosis /alopecia mucinosa**
3593. **✓Generalized myxedema (myxoedema)**
3594. **✓ Cutaneous Mucinosis of infancy CMI**
3595. **✓Papulonodular mucinosis associated with autoimmune connective tissue diseases**

3596. ✓papular mucinosis of toxic oil syndrome
3597. ✓Self-healing juvenile cutaneous Mucinosis
3598. ✓Digital mucous cyst
3599. ✓Trauma-induced cutaneous mucinosis
3600. ✓Lichen myxoedematosus
- ✓Nodular form
- ✓Discrete papular
- ✓Acral persistent papular

3601. ✓ Reticular erythematous mucinosis REM
3602. ✓ Nevus mucinosus
3603. ✓Neuropathia mucinosa cutanea
3604. ✓Nevoid follicular mucinosis
3605. ✓Obesity associated lymphedematous mucinosis
3606. ✓Postirradiation localized mucinosis
3607. ✓Pretibial myxedema (myxoedema)
3608. ✓Scleredema
3609. ✓Scleredema of buschke=Scleredema diabeticorum
3610. ✓Self-healing cutaneous mucinosis
3611. ✓Urticaria-like follicular mucinosis
3612. Mucinous carcinoma
3613. Mucinous syringometaplasia
3614. Muckle–wells syndrome
3615. Mucocele
3616. Mucoepidermoid carcinoma
3617. Mucolipidosis
3618. **Mucopolysaccharidosis**
3619. ✓Hurler-scheie syndrome (mps i-h=s)
3620. ✓Hurler/Scheie
3621. ✓ scheie syndrome (mps is)
3622. ✓ Hunter syndrome (mps ii)
3623. ✓sanfilippo syndrome (mps iii):Type A ,B,C,D

3624. ✓ morquio syndrome (mp iv):Type A ,B
3625. ✓ maroteaux-lamy syndrome (mps vi)
3626. ✓ sly syndrome (mps vii)
3627. Mucosal neuroma
3628. Mucositis
3629. Mucormycosis
3630. Mudichood
3631. Muir- torre syndrome
3632. Mukamel syndrome
3633. Mulvihill–smith syndrome
3634. Mullerian cyst
3635. Mumps
3636. Multifocal lymphangioendotheliomatosis with thrombocytopenia= cutaneovisceral angiomatosis with thrombocytopenia
3637. Multicentric osteolysis, nodulosis, and arthropathy MONA
3638. Multiple cartilaginous exostosis Diaphyseal aclasis
3639. Multiple circumscribed lipomas
3640. Multiple cutaneous linear neuromas (MCLN)
3641. Multiple endocrine neoplasia type MEN 1: wermer
3642. Multiple endocrine neoplasia type 2 (MEN 2A, sipple syndrome)
3643. Multiple endocrine neoplasia MEN type 3 (MEN 2B)= Multiple mucosal neuroma syndrome
3644. Multiple hamartoma syndrome
3645. Multiple hemangiomas of infancy
3646. Multiple minute digitate hyperkeratosis
3647. Multiple myeloma
3648. Multinucleate cell angiohistiocytoma
3649. Multiple pterygia= escobar syndrome
3650. Multiple self-healing palmoplantar carcinoma
3651. Multiple sulfatase deficiency (mucosulfatidosis)
3652. Munchausen syndrome

3653. **Mycetoma & eumycotic mycetoma & actinomycotic mycetoma & madura foot& maduromycosis**
3654. **Mycoplasma pneumoniae-induced rash and mucositis (MIRM)**
3655. **Myelodysplastic syndrome**
3656. **Myeloid sarcoma**
3657. **Myeloperoxidase deficiency**
3658. **Myeloproliferative disorders :Essential thrombocythemia**
3659. **Myoepithelioma**
3660. **Myoepithelial carcinoma**
3661. **Myopericytoma**
3662. **Myhre syndrome**
3663. **Cutaneous Myiasis(Furuncular Myiasis&Migratory Myiasis (Creeping Myiasis)&Wound Myiasis)**
3664. **Cavitary Myiasis (Nasal& aural&Ocular&Genitourinary)**
3665. **Accidental Myiasis (Pseudomyiasis)**
3666. **Nosocomial Myiasis**
3667. **Myointimoma**
3668. **Myofibromatosis (Adult & infantile)**
3669. **Myofibroblastic sarcoma=Myofibrosarcoma**
3670. **Inflammatory myofibroblastic tumor =Cutaneous inflammatory pseudotumor**
3671. **Myolipoma**
3672. **Myonecrosis**
3673. **Myotonia**
3674. **Myotonic dystrophy**
3675. **Myxofibrosarcoma**
3676. **Myxoinflammatory fibroblastic sarcoma**
3677. **Myxoid cyst**
3678. **Naegeli-franceschetti-jadassohn syndrome**
3679. **Nail dystrophy–deafness syndrome**
3680. **Nakajo–nishimura syndrome**
3681. **NAME syndrome ((nevi, atrial myxoma, mucinosis of the skin and ephelides syndrome)**

3682. Napkin dermatitis (primary irritant contact napkin dermatitis)
3683. Allergic contact napkin dermatitis
3684. Nasolabial cyst
3685. Nasopalatine duct cyst =cyst of the incisive papilla
3686. Natal teeth
3687. Natal teeth, patent ductus arteriosus, intestinal pseudo-obstruction
3688. **Navaratnas in Dermatology(see next)**
3689. ✓Cat eye syndrome
3690. ✓ Coral bead sign
3691. ✓Coral dermatitis
3692. ✓Coral reef bleaching
3693. ✓Coral red fluorescence
3694. ✓ Diamond finger gap test
3695. ✓Diamond hair pattern
3696. ✓ Diamond peel
3697. ✓Diamond necklace appearance
3698. ✓ Diamond's medium
3699. ✓Diamond keratinocytes
3700. ✓Diamond palmar pattern
3701. ✓ Diamond-shaped depigmented patch (forehead)
3702. ✓ Diamond scales
3703. ✓ Diamond skin disease
3704. ✓ Diamond's triad
3705. ✓ Emerald laser
3706. ✓Epstein pearls
3707. ✓ Gardner-Diamond syndrome
3708. ✓Horn pearls or keratin pearls
3709. ✓ Pearl fractional laser
3710. ✓Pearly penile papules
3711. ✓Pearly wreath
3712. ✓Red coral vessels

3713. ✓ Ruby laser
3714. ✓ Ruby spots
3715. ✓Smegma pearls
3716. ✓String of pearls
3717. ✓Sapphire lasers
3718. Necrobiosis lipoidica
3719. Necrobiotic xanthogranuloma
3720. Necrolytic acral erythema
3721. Necrolytic migratory erythema
3722. Necrotizing eosinophilic folliculitis
3723. Necrotizing fasciitis= flesh eating bacteria syndrome
3724. Necrotizing infundibular crystalline folliculitis
3725. Necrotizing lymphocytic folliculitis
3726. Necrotizing neutrophilic dermatosis
3727. Necrotizing sialometaplasia
3728. Neonatal progeroid syndrome=weidemann-rautenstrauch
3729. Neonatal cephalic pustulosis
3730. Neonatal cold injury
3731. Neonatal impetigo
3732. Neonatal inflammatory skin and bowel disease 1 (NISBD1)
3733. Neonatal Onset Multi-Inflammatory disease NOMID
3734. Neonatal varicella
3735. Nephrogenic systemic fibrosis
3736. Nephrogenic fibrosing dermopathy
3737. Néstor–guillermo progeria syndrome
3738. NERD: nodules, eosinophilia, rheumatism, dermatitis and swelling
3739. Nerve sheath myxoma
3740. Peripheral nerve sheath tumor
3741. Malignant nerve sheath tumors= neurosarcoma = malignant schwannoma
3742. Neu laxova syndrome
3743. Neural tube defects
3744. Neuroblastoma

3745. **Neurocristic hamartoma**
3746. **Neurocutaneous (leptomeningeal) melanocytosis**
3747. **Neuroendocrine tumours**
3748. **Neurodermatitis**
3749. **Circumscribed neurodermatitis**
3750. **Neurofibroma**
3751. **Plexiform neurofibroma**
3752. **Neurofibromatosis (see next)**
3753. **✓Nf1 von recklinghausen disease**
3754. **✓Nf2 acoustic schwannoma**
3755. **✓Nf3 * mixed (central and peripheral neurofibromas)**
3756. **✓Nf4 * variant**
3757. **✓Nf5 * ,† segmental café-au-lait macules, neurofibromas, or both**
3758. **✓Nf6 * ,‡ café-au-lait macules only**
3759. **✓Nf7 * late onset**
3760. **✓Nf-nos not otherwise specified**
3761. **Neurofibrosarcoma**
3762. **Neurofollicular hamartoma=spindle cell predominant trichodiscoma**
3763. **Neurolipomatosis=neural fibrolipoma= fibrolipomatous hamartoma of nerve**
3764. **Traumatic Neuroma**
3765. **Palisaded encapsulated neuroma= Solitary Circumscribed Neuroma**
3766. **Epithelial sheath neuroma**
3767. **Neurosarcoma**
3768. **Neurothekeoma=Dermal nerve sheath myxoma**
3769. **Atypical cellular neurothekeoma**
3770. **Neurotic excoriation= dermatillomania =skin-picking disorder**
3771. **Neutral lipid storage disease**
3772. **✓Severe congenital neutropenia**
3773. **✓Cyclic neutropenia**
3774. **✓X-linked neutropenia**
3775. **✓Poikiloderma with neutropenia, clericuzio type**

3776. **Neutrophilic dermatosis of dorsal hands= Pustular Vasculitis**
3777. **Neutrophilic eccrine hidradenitis NEH**
3778. **Neutrophilic figurate erythema**
3779. **Neutrophilic sebaceous adenitis**
3780. **Neutrophilic superficial eccrine ductitis**
3781. **Neutrophilic urticarial dermatosis**
3782. **Nevada syndrome (nevus epidermicus verrucosus with angiodysplasia and aneurysms)**
3783. **Nevoid acanthosis nigricans=rounded and velvety epidermal nevus RAVEN**
3784. **Nevoid basal cell carcinoma syndrome=Gorlin Goltz syndrome= Basal cell nevus syndrome**
3785. **Nevoid hamartoma**
3786. **Nevoid hypertrichosis**
3787. **Nevoid melanoma**
3788. **Acquired Nevus (juncional & compound & Intradermal)=Nevocellular Nevus= mole=Cellular Nevus**
3789. **Agminated Nevus**
3790. **Ancient nevus**
3791. **Nevus anelasticus**
3792. **Nevus anemicus- pharmacologic nevus**
3793. **Apocrine nevus**
3794. **Naevus araneus**
3795. **Becker melanosis/nevus=pigmented Hairy epidermal Nevus**
3796. **Becker naevus syndrome**
3797. **Blitz nevus**
3798. **Blue nevus (SEE next)**
3799. **✓Common**
3800. **✓Cellular:Typical & Atypical**
3801. **✓ Epithelioid**
3802. **✓Malignant =blue nevus like melanoma**
3803. **✓Target**
3804. **✓Patch like**

3805. ✓Plaque like
3806. ✓Large plaque-type blue naevus with subcutaneous cellular nodules (LPTBN-SN)
3807. ✓Hypochromic=Amelanotic
3808. ✓Agminate
3809. ✓compound
3810. ✓Sclerosing
3811. ✓Epithelioid and fusiform blue nevus of chronically sun-damaged skin
3812. Nevus brauer
3813. Cerebriform nevus
3814. Nevus cesius (segmental dermal melanocytosis)
3815. Combined Nevus = biphasic melanocytic nevus
3816. Nevus comedonicus
3817. Nevus comedonicus syndrome
3818. Congenital Nevus (small & medium& large & giant)
3819. Congenital melanocytic nevus (CMN)=nevus pigmentosus et pilosus=Nevus pilosus
3820. Bathing trunk nevus =Giant pigmented nevus
3821. Congenital panfollicular nevus
3822. Connective tissue nevus
3823. Botryoid connective tissue nevus
3824. Combined epidermal-connective tissue nevus of proteoglycan
3825. Nevus en cocarde - cockade
3826. Nevus corniculatus
3827. Nevus corymbosa
3828. Deep Penetrating nevus
3829. Nevus depigmentosa= achromic nevus
3830. Desmoplastic nevus
3831. Divided nevus
3832. Divided nevus of the eyelid (panda)
3833. Divided nevus of the penis
3834. Dysplastic nevus=clark nevus= Atypical nevus

3835. Eccrine nevus
3836. Mucinous Eccrine nevus MEN
3837. Eccrine Nevus of the Castori Type
3838. Nevus eclipse
3839. Nevus of epidermolysis bullosa
3840. Epidermal Nevus EN
3841. Epidermolytic Epidermal Nevus
3842. Keratinocytic Epidermal Nevus
3843. Epidermal Verrucous Nevus
3844. Nevus unius lateralis
3845. Epidermal naevus – Hailey–Hailey type
3846. Epidermal naevus = Darier type = Acantholytic dyskeratotic epidermal nevus
3847. Epidermal nevus syndrome= Schimmelpenning–feuerstein–mims syndrome
3848. Papular epidermal naevus with ‘‘skyline’’ basal cell layer (PENS)
3849. Fibroblast growth factor receptor 3 epidermal nevus syndrome (garcia-hafner-happle syndrome)
3850. Segmental seborrheic keratosis-like keratinocyte epidermal nevus
3851. Hystrix-like epidermal nevus of nevada syndrome
3852. Inflammatory linear verrucous epidermal nevus (ILVEN)= Dermatitic epidermal Nevus=eczematous epidermal Nevus
3853. Kerinokeratoticus nevus
3854. Glycosaminoglycan Nevi= nevus mucinosus
3855. Halo Nevus=leukoderma acquisitum centrifugum= sutton’s nevus
3856. Hori Nevus(acquired, bilateral nevus of ota- like macules)
3857. Nevus of ito=nevus fuscoceruleus acromiodeltoideus
3858. Naevus Lentiginosus Linearis
3859. Nevus lipomatosis superficialis= hoffman-zurhalle nevus
3860. Melanocytic nevi of genital skin
3861. Melanocytic nevus of acral skin
3862. Miescher nevus
3863. Myerson nevus = Halo dermatitis=Halo eczema

3864. **Naevus marginatus**
3865. **Naevus oligaemicus**
3866. **Oral pigmented nevus**
3867. **Ota nevus =Nevus Fuscoceruleus Ophthalmomaxillaris**
3868. **Sun nevus= nevus of Ota acquisita**
3869. **Nevus osteo nanta**
3870. **Nevus psiloliparus**
3871. **Reactivated nevus**
3872. **Recurrent nevus= pseudomelanoma**
3873. **Rhodoid nevus =capillary malformation-arteriovenous malformation**
3874. **Nevus roseus**
3875. **Sclerosing nevus**
3876. **Nevus sebaceous of Jadassohn**
3877. **Nevus simplex & angel's kiss &stork bite &salmon patch**
3878. **Spark's nevus**
3879. **Nevus spilus=speckled lentiginous nevus=zosteriform lentiginous nevus(Papular Nevus Spilus & Macular Nevus Spilus)**
3880. **Papular Nevus Spilus syndrome**
3881. **Speckled lentiginous nevus syndrome**
3882. **Naevus spinofiliformis**
3883. **✓Spitz Nevus =juvenile melanoma (Agminated& Disseminated)**
3884. **✓Atypical Spitz tumors**
3885. **✓Malignant =metastasizing Spitz tumors**
3886. **Pigmented spindle cell nevus of reed**
3887. **Spotted grouped pigmented nevi =eccrine centered nevus**
3888. **Nevus of striated muscle**
3889. **Nevus sudiferous**
3890. **Targetoid hemosiderotic nevus**
3891. **Nevus tricholemmocycticus**
3892. **Unna nevus = papillated dermal nevus**
3893. **Large venous nevus**
3894. **Small venous nevi**

3895. **Nevus venous of servelle- martorell type**
3896. **Nevus vascularis mixtus (cutaneous vascular twin nevi)**
3897. **Nevus vascularis mixtus, a hallmark of mixed vascular nevus syndrome (ruggieri- leech syndrome)**
3898. **Nevus xanthoma**
3899. **Newborn exfoliation= physiological desquamation**
3900. **Newborn and neonatal reactions & diseases**
3901. **Nezelof syndrome**
3902. **Nicolaides–baraitser syndrome**
3903. **Nicolau syndrome**
3904. **Nicotine stomatitis**
3905. **Niemann–pick disease**
3906. **Nijmegen breakage syndrome (NBS)**
3907. **Nipple adenoma**
3908. **Chronic nipple eczema**
3909. **Absent, hypoplastic or anomalous nipples**
3910. **Nevoid hyperkeratosis of nipple**
3911. **Nlrp1-associated autoinflammation with arthritis and dyskeratosis**
3912. **Monogenic nod-like receptor family pyrin domain containing 1 (NLRP1)-associated diseases**
3913. **Nocardiosis**
3914. **Nodular colloid degeneration**
3915. **Nodular elastosis**
3916. **Nodular fasciitis=nodular pseudosarcomatous fasciitis**
3917. **Nodular scleroderma**
3918. **Nodular vasculitis**
3919. **Nonne-milroy-meige syndrome**
3920. **Noonan syndrome**
3921. **Nonsuicidal Self-Injury**
3922. **Nontropical diabetic hand syndrome**
3923. **Notalgia parasethica**
3924. **Nuchal-type fibroma**
3925. **Nuchal fibrocartilaginous pseudotumor**

3926. **Nummular dermatitis**
3927. **Nucleotide-binding oligomerization domain (nod)-like receptor family pyrin domain containing 12 (nlrp12)-associated disorder (nlrp12ad)**
3928. **Nuclear factor-kappa b essential modulator (NEMO) deficiency syndrome**
3929. **Oasthouse disease**
3930. **Obesity associated lymphedematous mucinosis**
3931. **Obsessive–compulsive disorders**
3932. **Occipital horn syndrome**
3933. **Ochre Dermatitis**
3934. **Ochronosis**
3935. **Lowe syndrome (oculo-cerebro-renal syndrome)**
3936. **Oculocerebral syndrome with hypopigmentation**
3937. **Oculocerebrocutaneous syndrome (delleman syndrome)**
3938. **Oculodental syndrome=rutherfurd syndrome**
3939. **Oculo-dento-digital dysplasia**
3940. **Oculodermal melanocytosis**
3941. **Oculoectodermal syndrome**
3942. **Oculofaciocardiodental (OFCD)**
3943. **Oculotrichodysplasia**
3944. **Odontogenic cysts**
3945. **Odonto tricho ungual digital palmar syndrome**
3946. **Odonto trichomelic syndrome**
3947. **Odontomicronychial dysplasia**
3948. **Odonto-onycho-dermal ectodermal dysplasia**
3949. **Odonto-onychodysplasia with alopecia**
3950. **Oleoma**
3951. **Oliver–mcfarlane syndrome**
3952. **Ollier dyschondroplasia**
3953. **Omenn syndrome**
3954. **Omphalomesenteric cyst**
3955. **Omphalomesenteric duct anomalies**
3956. **Omphalocoele**
3957. **Onchocerciasis =river blindness=erysipelas de la costa=sowda**

3958. **Onychotrichodysplasia and neutropenia,mental retardation**
3959. **Oncocytoma**
3960. **Oral allergy syndrome**
3961. **Oral cancrum oris =Noma**
3962. **Oral facial digital syndrome (SEE next)**
3963. **✓ type i: papillon-league-psaume syndrome**
3964. **✓type ii: mohr syndrome**
3965. **✓Type iii: sugarman syndrom**
3966. **✓type iv: baraitser-burn syndrome**
3967. **✓type v: thurston syndrome**
3968. **✓ type vi: varadi-papp syndrome**
3969. **✓ type vii: whelan syndrome**
3970. **✓ type viii: oral-facial-digital syndrome, edwards type)**
3971. **Oral focal mucinosis**
3972. **Oral melanoma**
3973. **Oral nevus**
3974. **Oral papiiloma**
3975. **Oral Plasma Cell Mucositis**
3976. **Oral submucous fibrosis**
3977. **Orange palpebral spots**
3978. **Orf = ecthyma contagiosum = contagious pustular dermatitis**
3979. **Ornithosis (psittacosis)**
3980. **Orofacial granulomatosis OFG**
3981. **Oro-oculo-genital syndrome**
3982. **Osler–rendu–weber disease**
3983. **Osler's nodes**
3984. **Osteochondroma**
3985. **Osteochondrolipoma**
3986. **Osteogenesis imperfect (8 types)**
3987. **Ossification**
3988. **Ossifying fibromyxoid tumor**
3989. **Ossifying lipoma**

3990. **Ossifying plexiform tumor**
3991. **Osteomata cutis**
3992. **Osteosarcoma**
3993. **Osteomyelitis**
3994. **Osteopoikilosis**
3995. **Oestrogen hypersensitivity vulvovaginitis**
3996. **Oto-onycho-peroneal syndrome**
3997. **otopalatodigital syndrome**
3998. **Otophyma**
3999. **Otulipenia= otulin-related autoinflammatory syndrome (oras)=otulipenia)**
4000. **Oudsthoorn disease**
4001. **p14 deficiency**
4002. **Pachyderma**
4003. **Pachydermatoglyphy**
4004. **Pachydermatous eosinophilic dermatitis**
4005. **Pachydermia oralis**
4006. **Pachydermodactyly**
4007. **Pachydermoperiostosis=Primary Hypertrophic Osteoarthropathy**
4008. **pachyonychia congenita**
4009. **Pachyonychia congenita with amyloidosis and hyperpigmentation**
4010. **Pachyonychia congenita with leukonychia**
4011. **Pacinian corpuscle hyperplasia/pacinian neuroma**
4012. **Paederus/ beetle dermatitis**
4013. **Paget diseases**
4014. **Extramammary paget's disease**
4015. **Pagetoid Dyskeratosis**
4016. **Pagetoid Dyskeratosis of the Scrotum**
4017. **Palatine papilla cyst**
4018. **Palatal exostosis**
4019. **Pallister–killian syndrome**
4020. **Palisaded encapsulated neuroma (PEN)**
4021. **Palisaded neutrophilic and granulomatous dermatitis PNGD**
4022. **Palpable migratory arciform erythema**

4023. **Palmar erythema**
4024. **Palmar fasciitis and polyarthritis syndrome**
4025. **Palmoplantar erythrodysesthesia syndrome**
4026. **Palmoplantar pustulosis**
4027. **Palmoplantar pustular psoriasis**
4028. **Palmoplantar and perioroficial keratoderma with corneal epithelial dysplasia**
4029. **Palmoplantar keratoderma (PPK)**
4030. **Focal palmoplantar keratodermas (ppk)(see next)**
4031. ✓ **Richner–hanhart syndrome=Oculocutaneous Tyrosinemia**
4032. ✓ **Carvajal Huerta syndrome**
4033. ✓ **Howel–Evans syndrome= Tylosis with esophageal cancer**
4034. ✓ **Pachyonychia congenita type I (Jadassohn–Lewandowsky)**
4035. ✓ **Pachyonychia congenita type II (Jackson–Sertoli)**
4036. ✓ **Pachyonychia congenita type III (Schafer–Branauer)**
4037. ✓ **Pachyonychia congenita type IV (Tarda)**
4038. ✓ **Hereditary keratosis palmoplantaris variant of Wachters**
4039. ✓ **PPK and woolly hair**
4040. ✓ **focal ppk, isolated: Brünauer–Fuhs–Siemens**
4041. ✓ **ppk areata type**
4042. ✓ **striate ppk type 1,type 2,type3**
4043. ✓ **focal palmoplantar and gingival keratosis**
4044. ✓ **Palmoplantar keratoderma-congenital alopecia 1, Stefanovic type =keratoderma-hypotrichosis-leukonychia totalis syndrome, KHLS)**
4045. ✓ **PPK and congenital alopecia 2, Wallistype (PPKCA2, cataract-alopecia-sclerodactyly syndrome, CASS)**
4046. **Punctate palmoplantar keratodermas (ppk) (see next)**
4047. ✓ **punctate ppk buschke–fischer–brauer (type1)=Keratosis palmoplantaris papulosa**
4048. ✓ **ppk spiny music box= punctate palmoplantar keratoderma type 2 = filiform hyperkeratosis= digitate keratosis**

4049. ✓Marginal papular keratoderma,acrokeratoelastoidosis, focal acral hyperkeratosis (type 3 punctate ppk)
4050. ✓punctate keratoses of the palmar creases
4051. ✓porokeratosis punctata palmaris et plantaris
4052. ✓Focal acral hyperkeratosis
4053. ✓Papuloverrucous palmoplantar keratoderma of Jakac-Wolf=polykeratosis of touraine
4054. ✓Cole disease
4055. ✓PLACK syndrome (peeling skin-leukonychia-acral punctate keratosis-cheilitis, and knuckle pad)
4056. **Diffuse palmoplantar keratodermas (ppk)(see next)**
4057. ✓ Nonepidermolytic PPK Unna-Thost
4058. ✓Vörner epidermolytic palmoplantar keratoderma
4059. ✓unna–Thost epidermolytic palmoplantar keratoderma
4060. ✓Mal de meleda
4061. ✓ Nagashima, kimonis
4062. ✓ Bothnia
4063. ✓Greither epidermolytic palmoplantar keratoderma
4064. ✓Palmoplantar keratoderma of Sybert
4065. ✓Hidrotic ectodermal dysplasia (Clouston) syndrome
4066. ✓KLICK :keratosis linearis, ichthyosis congenita, sclerosing keratoderma
4067. ✓ Bart–pumphrey syndrome=Knuckle pads, leukonychia and deafness
4068. ✓Naxos disease= Mal de Naxos
4069. ✓Schöpf–schulz–passarge syndrome
4070. ✓Haim–munk syndromes
4071. ✓ppk mitochondrial
4072. ✓ppk due to connexin 26 defect
4073. ✓Palmoplantar keratoderma with deafness
4074. ✓PPK with nail dystrophy, and motor and sensory neuropathy
4075. ✓Huriez =scleroatrophic and keratotic dermatosis of the limbs
4076. ✓Loricin keratoderma : Camisa variant of Vohwinkel's syndrome

4077. ✓Camisa syndrome
4078. ✓Clouston syndrome
4079. ✓Corneodermatoosseous syndrome
4080. ✓Norbotten
4081. ✓Olmsted syndrome
4082. ✓Papillon–lefèvre syndrome
4083. ✓Arrhythmogenic right ventricular dysplasia, with mild palmoplantar keratoderma and woolly hair
4084. ✓Diffuse epidermolytic palmoplantar keratoderma (EPPK)
4085. ✓Keratosis palmoplantaris papillomatosa et verrucosa Jakac–Wolf
4086. ✓Vohwinkel syndrome =Keratoma hereditarium mutilans
4087. ✓ppk with sex reversal
4088. ✓PPK with pigmentation defects and skin carcinoma
4089. ✓Hyperkeratosis-hyperpigmentation syndrome
4090. ✓Gamborg -Nielsen keratoderma
4091. ✓curly hair–acral keratoderma–caries syndrome
4092. Papillomatoverrucous palmoplantar keratoderma
4093. Keratoderma Associated with Internal Malignancy
4094. Aquagenic transient reactive papulotranslucent acrokeratoderma= Aquagenic syringeal acrokeratoderma
4095. Pangeria
4096. Panfolliculoma
4097. **Panniculitis (see next)**
4098. ✓ Panniculitis due to blunt trauma
4099. ✓ Alpha 1 antitrypsin deficiency panniculitis
4100. ✓ Cold panniculitis
4101. ✓ Cytophagic histiocytic panniculitis
4102. ✓ Factitial
4103. ✓ Pancreatic panniculitis=Enzymic panniculitis
4104. ✓ Post radiotherapy
4105. ✓ Post-steroid

4106. ✓ Rothmann-Makai
4107. ✓ Weber-christian= relapsing febrile nodular non-suppurative panniculitis syndrome
4108. ✓ Annular atrophic connective tissue panniculitis of the ankles
4109. ✓ Atrophic connective tissue panniculitis
4110. ✓ Atypical lymphocytic lobular panniculitis
4111. ✓ Crystal deposition panniculitis
4112. ✓ Cutaneous polyarteritis nodosa
4113. ✓ Equestrian panniculitis
4114. ✓ Erythema induratum (nodular vasculitis)
4115. ✓ Erythema nodosum
4116. ✓ Fasciitis–panniculitis syndrome
4117. ✓ Idiopathic lobular panniculitis of pregnancy
4118. ✓ Infantile-onset panniculitis with uveitis and systemic granulomatosis
4119. ✓ Infection-induced panniculitis
4120. ✓ Lipodermatosclerosis
4121. ✓ Lipodystrophic panniculitis
4122. ✓ Lipophagic panniculitis of childhood
4123. ✓ lupus erythematosus panniculitis
4124. ✓ Malignancy-related panniculitis-like infiltrates
4125. ✓ Morphea/scleroderma panniculitis
4126. ✓ Pancreatitis, panniculitis, and polyarthritis (PPP)
4127. ✓ Panniculitic bacterid
4128. ✓ Panniculitis of dermatomyositis
4129. ✓ Panniculitis due to other injectable substances
4130. ✓ Panniculitis of lyme disease
4131. ✓ Plasma cell panniculitis
4132. ✓ Sclerema neonatorum
4133. ✓ Subcutaneous fat necrosis of the newborn
4134. ✓ Subcutaneous panniculitis -like t -cell lymphoma
4135. ✓ Superficial thrombophlebitis

4136. **Pantoea (enterobacter) agglomerans**
4137. **PAC (pyoderma gangrenosum, acne and ulcerative colitis)**
4138. **PAPA syndrome (pyogenic arthritis, pyoderma gangrenosum and acne)**
4139. **PAPASH syndrome (pyogenic arthritis, acne, pyoderma gangrenosum and suppurative hidradenitis)**
4140. **PASH syndrome (pyoderma gangrenosum , acne and suppurative hidradenitis)**
4141. **PASS (pyoderma gangrenosum, acne, and ankylosing spondylitis, with or without suppurative hidradenitis)**
4142. **PSAPASH (psoriatic arthritis, pyoderma gangrenosum, acne and hidradenitis suppurativa)**
4143. **Papillary carcinoma**
4144. **Papillary eccrine adenoma=tubular apocrine adenoma**
4145. **Papillary endothelial hyperplasia=masson's tumor**
4146. **Papillary hemangioma**
4147. **Papillary hyperplasia of the palate**
4148. **Papillary intralymphatic angioendothelioma (pila)=dabska-type hemangioendothelioma=dabska tumor=endovascular papillary angioendothelioma**
4149. **Papillomatosis cutis carcinoides gottron**
4150. **Papillomatosis cutis lymphostatica**
4151. **Papillon–léage syndrome**
4152. **Papillon–léfèvre syndrome**
4153. **Papular Acantholytic Dyskeratosis**
4154. **Papular clear cell hyperplasia of the eccrine duct**
4155. **Papular dermatitis= chronic papular urticarial= Chronic prurigo of adults**
4156. **Papular epidermal nevus with skyline**
4157. **Papular eruption of blacks**
4158. **Papular purpuric gloves and socks syndrome**
4159. **Papuloerythroderma of ofuji**
4160. **Parachordoma**
4161. **Paracoccidioidomycosis=south american blastomycosis**
4162. **Paraffinoma**

4163. **Paraganglioma-like dermal melanocytic tumor**
4164. **Paraquat tongue**
4165. **Paragonimiasis**
4166. **Parapox**
4167. **Parakeratosis variegate**
4168. **parameatal urethral cyst**
4169. **Paraneoplastic erythroderma**
4170. **Paraneoplastic scleroderma-like syndrome**
4171. **Paraneoplastic pemphigus PNP**
4172. **Paraneoplastic autoimmune multiorgan syndrome PAMS**
4173. **Parapsoriasis :small plaque= chronic superficial scaly dermatitis (digitate dermatosis&xanthoerythrodermia perstans)**
4174. **Parapsoriasis :large plaque(parapsoriasis variegate =parakeratosis variegata =retiform parapsoriasis= parapsoriasis lichenoides &poikilodermatous Parapsoriasis)**
4175. **Parasitophobia**
4176. **Parinaud syndrome**
4177. **Parkes weber syndrome**
4178. **Paroxysmal nocturnal hemoglobinuria**
4179. **Parry-romberg syndrome**
4180. **Parulis**
4181. **Pasteurellosis**
4182. **pearly penile papules**
4183. **Pebbling**
4184. **PEComa (fibroma-like perivascular epithelioid cell tumor)**
4185. **pediculosis capitis**
4186. **Pediculosis corporis**
4187. **Pediculosis pubis & pthirus pubis=crab lice**
4188. **Phthiriasis palpebrarum**
4189. **Pediculid**
4190. **peeling skin syndromes**
4191. **Pellagra**
4192. **Perlman syndrome**

4193. **Pemphigus erythematous = senear-usher syndrome**
4194. **Erythema annulare centrifugum-like pemphigus erythematosus (EAC-LIKE PE)**
4195. **Pemphigoid gestationis =herpes gestation=gestational pemphigoid**
4196. **Pemphigus foliaceus**
4197. **Endemic pemphigus foliaceus= fogo selvage= brazilianpemphigus foliaceus**
4198. **Pemphigus herpetiformis**
4199. **IgA Pemphigus :intraepidermal neutrophilic type**
4200. **IgA Pemphigus : subcorneal pustular dermatosis**
4201. **Pemphigus syphiliticus**
4202. **Contact pemphigus**
4203. **Pemphigus vegetans(Hallopeau type & Neumann type)**
4204. **Pemphigus vulgaris**
4205. **Juvenile pemphigus vulgaris**
4206. **Verrucous pemphigus=pemphigus acanthomata=postpemphigus acanthoma**
4207. **Neonatal pemphigus vulgaris**
4208. **Pena-shokeir syndrome**
4209. **Pendulous fibroma**
4210. **Penicillamine-induced elastosis perforans**
4211. **Penicilliosis**
4212. **Penile acne**
4213. **Carcinoma of the penis**
4214. **Penile intraepithelial neoplasia**
4215. **Penile lymphedema**
4216. **Penile phimosis &paraphimosis**
4217. **Penile plasma cell (zoon's) balanitis**
4218. **Penile sclerosing lymphangitis**
4219. **Penttinen syndrome**
4220. **Perianal and vulvovaginal (perineal) streptococcal infection**
4221. **Perianal comedones**
4222. **Perianal streptococcal dermatitis**
4223. **Periarticular thenar erythema with onycholysis (PATEO)**

4224. **Periflexural exanthema**
4225. **Perifollicular elastolysis**
4226. **Perifollicular fibroma**
4227. **Perinatal gangrene of the buttock**
4228. **Recurrent toxin-mediated (perineal) erythema**
4229. **Perineurioma=storiform perineurial fibroma=localized hypertrophic neuropathy=storiform perineurial fibroma=fibrolamellar nerve sheath tumor**
4230. **Malignant perineurioma**
4231. **Periodontitis**
4232. **Periodontal fistula**
4233. **Periodic fever with aphtous stomatitis, pharyngitis and adenitis syndrome**
4234. **Periodic fever, immunodeficiency, and thrombocytopenia**
4235. **Perioral dermatitis**
4236. **Periobital acquired dermal macular hyperpigmentation**
4237. **Periorificial dermatitis=childhood granulomatous periorificial dermatitis**
4238. **Peristomal skin related skin disorders**
4239. **Periumbilical calcific elastolysis**
4240. **Periumbilical perforating pseudoxanthoma elasticum**
4241. **Peroxisomopathies**
4242. **Peripheral giant cell granuloma**
4243. **Peripheral odontogenic fibroma**
4244. **Peripheral ossifying fibroma**
4245. **Periporitis staphylogenes = sweat gland abcess**
4246. **Pernio = chilblains**
4247. **Perniola syndrome**
4248. **Perlèche**
4249. **Petechiae**
4250. **Persistent cholinergic erythema**
4251. **Persistent acrovasculopathy syndrome**
4252. **Persistent Linear Bands in Infancy**
4253. **Peutz jeghers syndrome**
4254. **Peyronie’s disease**

4255. **Peyronellaea gardeniae infection**
4256. **PFAPA :periodic fever, aphthae, pharyngitis, and Adenitis syndrome**
4257. **Phalangeal sarcoma**
4258. **Phacomatosis phakomatosis (see next)**
4259. **✓Type i (adamson-best syndrome)**
4260. **✓Type ii (takano-kruger-doi syndrome)**
4261. **✓ Type iii (kobori-toda syndrome)**
4262. **✓Type iv**
4263. **✓ phakomatosis cesioflammea**
4264. **✓ phakomatosis spilorosea**
4265. **✓ phakomatosis melanovascularis**
4266. **✓ phakomatosis cesiomarmorata**
4267. **✓ phakomatosis pigmentovascularis**
4268. **✓ phakomatosis pigmentokeratotica**
4269. **Phakomatosis Pigmento-Eccrina**

Phenomena in dermatology (see next)

4270. **✓ alopecia areata (aa) sparing lesional gray/white hairs**
4271. **✓ anatopic phenomenon**
4272. **✓ atopic dermatitis generally spare the napkin area**
4273. **✓ butterfly sign**
4274. **✓ deck chair sign**
4275. **✓ ear lobe sign**
4276. **✓ irritant diaper dermatitis spares flexures**
4277. **✓ islands of sparing in pityriasis rubra pilaris**
4278. **✓ reverse koebner phenomenon**
4279. **✓ ichthyosis vulgaris spares flexures**
4280. **✓ epidermal growth factor receptor (egfr) inhibitor-associated rash spares irradiated areas**
4281. **✓ knuckle sparing of acute cutaneous lupus erythematosus (ACLE)**
4282. **✓ milian's ear sign**

4283. ✓ **nasolabial fold sparing in the malar rash of systemic lupus erythematosus**
4284. ✓ **nose sign**
4285. ✓ **paretic limbs spared in rheumatic diseases**
4286. ✓ **pityriasis rosea spare sun-exposed areas**
4287. ✓ **pruritic urticarial papules and plaques of pregnancy (puppp) spares periumbilical area**
4288. ✓ **psoriasis often spare central face**
4289. ✓ **relapsing polychondritis spare earlobes**
4290. ✓ **renbok phenomenon**
4291. ✓ **rosacea spares circum-oral and peri-orbital areas**
4292. ✓ **scrotum sparing in tinea cruris**
4293. ✓ **spared zones in leprosy**
4294. ✓ **sunbed suntan sacroscapular sparing sign**
4295. ✓ **upper axillary area spared in pityriasis versicolor**
4296. ✓ **white islands in a sea of red in dengue fever**
4297. ✓ **vermillion border spared in peri-oral dermatitis**
4298. ✓ **vermilion border spared in an acrodermatitis enteropathica (ae)-like syndrome**
4299. ✓ **wilkinson's triangle**
4300. ✓ **koebner reaction**
4301. ✓ **Isomosaictopic reaction**
4302. ✓ **wolf 's isotopic response**
4303. ✓ **Isovaccinetopic reaction**
4304. ✓ **Isoradiotopic reaction**
4305. ✓ **Isophototopic reaction**
4306. ✓ **Isotraumatopic reaction**
4307. ✓ **Isocaumatopic reaction**
4308. ✓ **Isoscartopic reaction**
4309. ✓ **Isotattotopic reaction:**
4310. ✓ **Isolymphtopic reaction**

4311. ✓ **Isovenostatic reaction**
4312. ✓ **Isoneuraltopic reaction**
4313. ✓ **Isoshockwave reaction**
4314. ✓ **Isodermatopic reaction**
4315. ✓ **reverse koebner reaction**
4316. ✓ **Isomosaictopic nonreaction**
4317. ✓ **wolf's isotopic nonreaction**
4318. ✓ **Isovaccinetopic nonreaction**
4319. ✓ **Isoradiotopic nonreaction**
4320. ✓ **Isophototopic nonreaction**
4321. ✓ **Isotraumatopic nonreaction**
4322. ✓ **Isocaumatopic nonreaction**
4323. ✓ **Isoscartopic nonreaction**
4324. ✓ **Isotattotopic nonreaction**
4325. ✓ **Isolymphtopic nonreaction**
4326. ✓ **Isovenostatic nonreaction**
4327. ✓ **Isoneuraltopic nonreaction**
4328. ✓ **Isoshockwave nonreaction**
4329. ✓ **Isodermatopic nonreaction**
4330. ✓ **Prozone phenomenon**
4331. ✓ **Pagetoid phenomenon**
4332. ✓ **Reynold's braude phenomenon**
4333. ✓ **Epitope spreading phenomenon**
4334. ✓ **Rebound phenomenon**
4335. ✓ **Sulzberger-chase phenomenon**
4336. ✓ **Harlequin phenomenon**
4337. ✓ **Splendore-hoeppli phenomenon**
4338. ✓ **Rumpel leede phenomenon**
4339. ✓ **Olfleck's phenomenon**
4340. ✓ **Ultraviolet recall phenomenon**
4341. ✓ **Raynaud's phenomenon**

4342. ✓ **Hoigne's phenomenon**
4343. ✓ **Bell's phenomenon**
4344. ✓ **Delayed blanch phenomenon**
4345. ✓ **Castling phenomenon**
4346. ✓ **Kasabach merritt phenomenon**
4347. ✓ **Halo phenomenon**
4348. ✓ **Lucio phenomenon**
4349. ✓ **Mauserung phenomenon**
4350. ✓ **Meyerson phenomenon**
4351. ✓ **Meirowsky phenomenon**
4352. ✓ **Brocq's phenomenon**
4353. ✓ **Pathergy phenomenon**
4354. ✓ **Nikolskiy's phenomenon**
4355. **Phenylketonuria PKU**
4356. **Phaeohyphomycosis**
4357. **Phlebolymphedema**
4358. **Phlebectasia**
4359. **Phosphaturic mesenchymal tumor**
4360. **Photoageing**
4361. **Photodamage**
4362. **Photodistributed eruptive telangiectasia**
4363. **photolocalized purpura**
4364. **Photosensitive spongiotic/lichenoid eruption of micropapules and plaques**
4365. **Phototoxic reaction**
4366. **Photoallergic reaction**
4367. **Photoleukomelanodermatitis of kobori**
4368. **Photo-onycholysis**
4369. **Phrynoderma**
4370. **Phylloid hypomelanosis**
4371. **Phylloid hypermelanosis**
4372. **Phylloid terminal hair nevus**
4373. **Physical purpura**

4374. **Physiological desquamation**
4375. **Physiologic hyperpigmentation**
4376. **Physiological melanoplakia**
4377. **Phytophotodermatitis**
4378. **PIBIDS (photosensitivity, Brittle nail , ichthyosis, brittle hair, intellectual impairment, decreased fertility and short stature)**
4379. **PBIDS(Photosensitivity-Brittle hair-Brittle nail-intellectual impairment-Decreased fertility-Short stature syndrome)**
4380. **Picker's nodule**
4381. **Piebaldism = white spot disease**
4382. **Black Piedra = Trichomycosis nodularis**
4383. **White piedra= Trichosporosis nodosa**
4384. **Pierre robin syndrome**
4385. **Piezogenic pedal papules**
4386. **Piezogenic wrist papules**
4387. **pigmented allergic salute sign**
4388. **Pigmented follicular cyst**
4389. **Pigmentary damarcation lines PDL= Futcher's lines=Voigt lines= Ito's lines**
4390. **Pigmentary mosaicism**
4391. **pigmentary twin spotting**
4392. **Pigmented macules of bony prominences (PMBP)**
4393. **Pigmented neuroectodermal tumour of infancy**
4394. **Pigmented purpura dermatosis ppd (see next)=purpura pigmentosa chronica**
4395. ✓ **Eczematid-like purpura of doucas and kapetanakis**
4396. ✓ **Lichen aureus=lichen purpuricus**
4397. ✓ **Familial pigmented purpuric eruption**
4398. ✓ **pigmented purpuric lichenoid dermatitis of gougerot and blum=lichenoid purpura = purpuric pigmented lichenoid dermatitis**
4399. ✓ **Linear pigmented purpura**
4400. ✓ **Granulomatous pigmented purpura**
4401. ✓ **Purpura annularis telangiectodes of majocchi**

4402. ✓ Schamberg disease =progressive pigmentary dermatosis
4403. pigmented transverse nasal band
4404. Pigmentatio reticularis faciei et colli
4405. Pik3ca-related segmental overgrowth syndrome
4406. Pilar cyst= wen =trichilemmal cyst= isthmus-catagen cyst
4407. Pilar sheath acanthoma
4408. Proliferating pilar tumor
4409. Proliferating trichilemmal cyst
4410. Malignant proliferating trichilemmal tumors
4411. Pili annulati
4412. Pseudopili annulati
4413. Pili bifuracti
4414. Pili migran
4415. Pili multigemini
4416. Pili torti
4417. Pili torti and developmental delay
4418. Pili torti and onychodysplasia (beare type)
4419. Pili torti and onychodysplasia
4420. pili trianguli =Uncombable hair=spun glass hair
4421. Pilodental dysplasia with refractive errors
4422. Pilomatricoma/ Pilomatrixoma= calcifying epithelioma of Malherbe
4423. Pilomatricomal horn
4424. Pilomatrical carcinoma
4425. Pilomatrixcarcinoma
4426. Pilonidal sinus
4427. Pinta
4428. Pintoid dyschromia of yaws
4429. Pipkin syndrome
4430. Pityriasis rubra pilaris PRP=lichen ruber pilaris = Lichen Ruber acuminatus
✓Classic adult
✓Atypical adult
✓Classic juvenile

- ✓ **Circumscribed juvenile**
- ✓ **Atypical juvenile**
- ✓ **Human immunodeficiency virus infection associated**

4431. **Pityriasis alba= impetigo pityroides= Pityriasis simplex= pityriasis maculata**
4432. **Pityriasis folliculorum**
4433. **Pityriasis lichenoid chronica PLC**
4434. **Pityriasis lichenoides et varioliformis acuta PLEVA**
4435. **Febrile ulceronecrotic variant of PLEVA =Febrile ulceronecrotic mucha-habermann disease FUHMD =Pityriasis lichenoides with Ulceronecrosis and Hyperthermia**
4436. **Pityriasis rosea (classic & vesicular & urticarial & target like& purpuric & pustular & abortive&inverted)**
4437. **pityriasis rosea Gibert**
4438. **Pityriasis rosea gigantea of darier**
4439. **Pityriasis circinata et marginata of widal**
4440. **Pityriasis rotunda= Pityriasis Circinata**
4441. **Pityriasis Steatoides**
4442. **Pitted keratolysis = keratolysis plantare sulcatum**
4443. **Pfeiffer syndrome**
4444. **Plague**
4445. **PLAID immune plcg2-associated antibody deficiency and immune dysregulation**
4446. **APLAID (autoinflammation and plcg2-associated antibody deficiency and immune dysregulation)**
4447. **Plantar fibromatosis = ledderhose disease**
4448. **Plaque-like CD34-positive dermal fibroma**
4449. **Plate-like osteoma cutis**
4450. **Plasma cell gingivitis =allergic gingivostomatitis= idiopathic gingivostomatitis= atypical gingivostomatitis**
4451. **Plasma cell glossitis**
4452. **Plasma cell granuloma**
4453. **Plasmacytoma**

4454. **Plasmacytosis**
4455. **Plasmoacanthoma**
4456. **Pleomorphic adenoma**
4457. **Carcinoma ex pleomorphic adenoma**
4458. **Pleomorphic dermal sarcoma**
4459. **Pleomorphic fibroma of the skin**
4460. **pleomorphic hyalinizing angiectatic tumor**
4461. **Pleomorphic lipoma**
4462. **Pleomorphic liposarcoma**
4463. **Pleomorphic Sarcoma**
4464. **Undifferentiated pleomorphic sarcoma**
4465. **Pleonosteosis**
4466. **Plexiform fibrohistiocytic tumor**
4467. **Plica polonica=neuropathica**
4468. **Plummer-vinson syndrome**
4469. **Pneumocystis carinii=PNEUMOCYSTOSIS**
4470. **Podoconiosis**
4471. **Poems syndrome (polyneuropathy, organomegaly, endocrinopathy, M-protein and skin changes)= crow-fukase syndrome**
4472. **Pohl–Pinkus constriction**
4473. **Poikiloderma**
4474. **Poikiloderma vasculare atrophicans**
4475. **Poikiloderma-like cutaneous amyloidosis**
4476. **Poikiloderma of civatte**
4477. **Poikilodermatous plaque-like hemangioma (PPLH)**
4478. **Pointlist melanotic macules**
4479. **Poison ivy dermatitis**
4480. **Poland syndrome**
4481. **Poliosis**
4482. **Pomade crust**
4483. **polyarteritis nodosa =panarteritis nodosa PAN**
4484. **Polycythemia rubra vera=Erythremia**
4485. **Polycystic ovarian syndrome (PCOS)**

4486. **Polymastia**
4487. **Polymorphous cold eruption**
4488. **Polymorphous light eruption PMLE**
4489. **polymorphous sweat gland carcinoma**
4490. **Polymorphous (low-grade) adenocarcinoma**
4491. **Polymyositis**
4492. **Polyostotic fibrous dysplasia**
4493. **Polythelia pilosa**
4494. **Pompholyx**
4495. **pool palms/pool toes**
4496. **Popliteal pterygium syndrome= klein syndrome**
4497. **Popsicle panniculitis**
4498. **Porokeratosis (see next)**
4499. **✓Porokeratosis of mibelli**
4500. **✓Disseminated superficial actinic porokeratosis (DSAP)**
4501. **✓ linear porokeratosis**
4502. **✓punctate porokeratosis**
4503. **✓Eruptive disseminated porokeratosis**
4504. **✓Bullous Porokeratosis**
4505. **✓Porokeratosis palmaris et plantaris et disseminata PPPD**
4506. **✓Eruptive pruritic papular porokeratosis**
4507. **✓porokeratosis ptychotropica**
4508. **✓Postirradiation multiple minute digitate porokeratosis**
4509. **✓Verrucous porokeratosis**
4510. **✓porokeratotic eccrine ostial and dermal duct nevus (PEODDN)**
4511. **✓Porokeratotic eccrine and hair follicle nevus PEHFN**
4512. **✓Porokeratotic Adnexal Ostial Nevus (PAON) = PEHFN and PEODDN**
4513. **✓Porokeratoma=porokeratotic acanthoma**
4514. **Linear palmo-plantar porokeratotic hamartoma**
4515. **Poroma**
4516. **Porocarcinoma**

4517. **Porphyria (see next)**
4518. ✓ **ALA dehydratase deficiency**
4519. ✓ **Acute intermittent porphyria**
4520. ✓ **Congenital erythropoietic porphyria= gunther syndrome CEP**
4521. ✓ **Hereditary coproporphyria**
4522. ✓ **Porphyria cutaneous tarda PCT**
4523. ✓ **Erythropoietic protoporphyria EPP**
4524. ✓ **Hepatoerythropoietic porphyria**
4525. ✓ **Pseudoporphyria= PCT-like syndrome=bullous dermatosis of hemodialysis=drug-induced pseudoporphyria cutanea tarda**
4526. ✓ **x-linked dominant protoporphyria**
4527. ✓ **Porphyria variegate=South African porphyria**
4528. **Portwine stain=nevus flammeus**
4529. **Postcardiotomy syndrome**
4530. **Post ECG leads dermatitis**
4531. **Post herpetic neuralgia**
4532. **Post inflammatory hyperpigmentation/postinflammatory melanosis**
4533. **Post inflammatory hypopigmentation/postinflammatory leukoderma**
4534. **Postirradiation digitate keratosis (PIDK)**
4535. **Post-irradiation pseudo-sclerodermatous panniculitis**
4536. **Post menopausal symptoms**
4537. **Post-miliarial exfoliation= post-heat rash exfoliation**
4538. **Post-stripping cutaneous sclerosis**
4539. **Post-thermal burn pruritus**
4540. **Post traumatic hematoma**
4541. **Powassan virus related skin infection**
4542. **Pipkin syndrome**
4543. **Prader willi syndrome**
4544. **Prayer nodule**
4545. **preauricular sinus**
4546. **Precalcaneal congenital fibrolipomatous hamartoma**

4547. **Pre- and postnatal growth retardation, mental retardation, and acral limb deficiencies**
4548. **Prepubertal Periodontitis**
4549. **Prepubertal unilateral fibrous hyperplasia of the labium majus**
4550. **Prepubertal vulval fibroma=childhood asymmetric labium majus enlargement**

Pregnancy diseases (see next)

4551. ✓**Atopic eruption of pregnancy = prurigo of pregnancy=prurigo gestationis (besnier) =early-onset prurigo of pregnancy = papular dermatitis of pregnancy = pruritic folliculitis of pregnancy = linear igm disease of pregnancy = eczema in pregnancy**
4552. ✓**Impetigo herpetiformis= pustular psoriasis of pregnancy**
4553. ✓**Pupp (pruritic urticarial papules and plaques of pregnancy= toxic erythema of pregnancy=late-onset prurigo of pregnancy = polymorphic eruption of pregnancy = toxemic rash of pregnancy**
4554. ✓**Intrahepatic cholestasis of pregnancy = Obstetric Cholestasis**
4555. ✓**prurigo gravidarum**
4556. ✓**Pregnancy-Related Pyogenic Granuloma (Granuloma Gravidarum)**
4557. ✓**Other physiologic changes**
4558. **premature gray/grey hair**
4559. **Pressure purpura (cough &vomiting)**
4560. **Pretibial angioplasia**
4561. **Primary signet-ring cell/histiocytoid carcinoma**
4562. **Primary cutaneous adenoid cystic carcinoma**
4563. **Primary cutaneous adenomyoepithelioma (pcAME)**
4564. **Primary cutaneous angioplasmacellular hyperplasia**
4565. **Primary cutaneous apocrine carcinomas / adenocarcinoma**
4566. **Primary cutaneous cribriform carcinoma (PCCC)**
4567. **Primary cutaneous extraskeletal osteosarcoma**
4568. **Primary lymphoepithelioma-like carcinoma of the skin (LELCS)**
4569. **Primary mucinous carcinoma of the skin**
4570. **Primary Thrombotic Microangiopathy**

4571. **Proctalgia fugax**
4572. **Autoimmune progesterone dermatitis AIPD**
4573. **Progesterone-induced stomatitis**
4574. **Werner's syndrome=progeria of the adult=pangeria**
4575. **Progeroid syndrome, petty type**
4576. **Hutchinson–Gilford progeria syndrome =progeria Infantum**
4577. **progressive cribriform and zosteriform hyperpigmentation PCZH**
4578. **Progressive macular hypomelanosis PMH**
4579. **Progressive osseus heteroplasia**
4580. **Progressive papular purpuric eruption**
4581. **Progressive Zosteriform Macular Pigmented Lesion (PZMPL)**
4582. **Prolidase deficiency(PD)=hyperimidodipeptiduria**
4583. **Proliferating pilar tumor**
4584. **Proliferative fasciitis**
4585. **Propionic acidemia**
4586. **Proteasome- associated autoinflammatory syndrome (PRAAS)**
4587. **Protein contact dermatitis**
4588. **Protein C deficiency**
4589. **Protein S deficiency**
4590. **Proteus syndrome= cerebriform fibrous proliferation**
4591. **Elattoproteus syndrome**
4592. **Protothecosis**
4593. **Prune belly syndrome**
4594. **Prurigiform angiomatosis (PA)**
4595. **Prurigo = Acne urticata**
4596. **Prurigo besnier**
4597. **Prurigo hebra**
4598. **Prurigo gravidarum**
4599. **Prurigo nodulris**
4600. **Prurigo pigmentosa**
4601. **prurigo of pregnancy**
4602. **Prurigo simplex**
4603. **Actinic prurigo = hutchinson's summer prurigo =hydroa aestivale**

4604. **Pruritic and dyskeratotic dermatosis**
4605. **Pruritus in scars/Keliod**
4606. **Aquagenic Pruritus**
4607. **Contagious Pruritus**
4608. **Pruritus ani**
4609. **Pruritus scroti**
4610. **Pruritus vulvae**
4611. **Renal Pruritus**
4612. **Scalp Pruritus**
4613. **Hematologic Pruritus**
4614. **Postmenopausal Pruritus**
4615. **Pruritus in Internal Malignancy**
4616. **Cholestatic Pruritus**
4617. **Senile Pruritus**
4618. **Endocrine pruritus**
4619. **Referred Pruritus (Mitempfindung)**
4620. **Psychogenic pruritus= Somatoform Pruritus**
4621. **Pharmacologic pruritus**
4622. **Pseudallescheriasis**

Pseudo in dermatology (see next)

4623. ✓ **Pseudo hypoparathyroidism**
4624. ✓ **Pseudo pseudo hypoparathyroidism**
4625. ✓ **Pseudoichthyosis, acquired**
4626. ✓ **Eruptive pseudoangiomatosis**
4627. ✓ **Pseudo acanthosis nigricans**
4628. ✓ **Pseudo acne of nasal crease**
4629. ✓ **Pseudo ainhum**
4630. ✓ **Pseudo albinism**
4631. ✓ **Pseudo alopecia areata**
4632. ✓ **Pseudo behcet's disease**
4633. ✓ **Pseudo blue-lunula**
4634. ✓ **Pseudo botryomycosis**

4635. ✓ **Pseudo bowen's disease**
4636. ✓ **Pseudo bubo**
4637. ✓ **Pseudo chancre redux**
4638. ✓ **Pseudo chromhidrosis**
4639. ✓ **Pseudo darier's sign**
4640. ✓ **Pseudo dermatomyositis**
4641. ✓ **Pseudo furuncle**
4642. ✓ **Pseudoglucagonoma**
4643. ✓ **Pseudo gout**
4644. ✓ **Pseudo halo nevus**
4645. ✓ **Pseudo horn cyst**
4646. ✓ **Pseudo hutchison's sign**
4647. ✓ **Pseudo lupus vulgaris**
4648. ✓ **Pseudo melanoma**
4649. ✓ **Pseudo monilethrix**
4650. ✓ **Pseudo mycetoma**
4651. ✓ **Pseudo mycosis fungoides**
4652. ✓ **Pseudo ochronosis**
4653. ✓ **Pseudo rheumatoid nodule**
4654. ✓ **Pseudosarcoma**
4655. ✓ **Pseudo scabies**
4656. ✓ **Pseudo scleroderma**
4657. ✓ **Pseudo sign of leser trelat**
4658. ✓ **Pseudo sporotrichosis**
4659. ✓ **Pseudoacromegaly**
4660. ✓ **Pseudocellulitis**
4661. ✓ **Pseudocyst of the auricle**
4662. ✓ **Pseudodidymosis aplasticosebacea**
4663. ✓ **Pseudoepitheliomatous hyperplasia**
4664. ✓ **Pseudofolliculitis barbae =pili recurvate= pili incarnati**
4665. ✓ **Pseudofolliculitis corporis**

4666. ✓ **Pseudofolliculitis vibrissae**
4667. ✓ **Pseudomamma**
4668. ✓ **Pseudomyogenic hemangioendothelioma**
4669. ✓ **Pseudomelanocytic nests**
4670. ✓ **Pseudopelade of brocq**
4671. ✓ **Pseudopili annulati**
4672. ✓ **Pseudotumor cerebri**
4673. ✓ **Pseudo kaposi**
4674. ✓ **Pseudo knuckle pad (chewing pad)**
4675. ✓ **Pseudo-koenen tumors**
4676. ✓ **Pseudomacrolunula**
4677. ✓ **Pseudo-pseudoxanthoma elasticum**
4678. ✓ **Pseudo-pseudo hutchison's sign**
4679. ✓ **Pseudovasculitis**
4680. **Pseudoepitheliomatous keratotic and micaceous balanitis**
4681. **Pseudoverrucous papule and nodule**
4682. **Perianal pseudoverrucous papules and nodules (PPPN)**
4683. **Pseudoxanthoma elasticum PXE= Grönblad-strandberg syndrome= systematized elastorrhexis**
4684. **Pseudoxanthoma elasticum-like papillary dermal elastolysis**
4685. **Pseudomonaus related diseases (see next)**
4686. ✓ **Pseudomonas hand foot syndrome**
4687. ✓ **Ecthyma gangrenosum**
4688. ✓ **Green nail syndrome**
4689. ✓ **Pseudomonal pyoderma**
4690. ✓ **Blastomycosis-like pyoderma**
4691. ✓ **Otitis externa**
4692. ✓ **pseudomonal folliculitis -hot tub folliculitis**
4693. ✓ **Malignant otitis externa**
4694. ✓ **Pseudomona chondritis**
4695. ✓ **Septicemia**

4696. **Psoriasis vulgaris=Chronic Plaque Psoriasis**
4697. **Annular Psoriasis**
4698. **Elephantine Psoriasis**
4699. **Erythrodermic Psoriasis**
4700. **Follicular Psoriasis**
4701. **Guttate Psoriasis**
4702. **Inverse Psoriasis**
4703. **Lichenoid psoriasis**
4704. **Linear Psoriasis**
4705. **Ostraceous Psoriasis**
4706. **Psoriasis :acrodermatitis continua of hallopeau**
4707. **Psoriasiform acral dermatitis**
4708. **Psoriasiform alopecia**
4709. **Psoriasiform keratosis**
4710. **Psoriasiform napkin dermatitis**
4711. **Psoriasis alba**
4712. **Psoriasis herpeticum**
4713. **Psoriasis punctate= micro-guttate**
4714. **Psoriasis vaccinalis**
4715. **Psoriatic arthritis**
4716. **Psoriatic neurodermiformis**
4717. **Psoriatic onycho-pachydermo-periostitis (POPP)**
4718. **Pustular Psoriasis**
4719. **Generalized pustular psoriasis of pregnancy**
4720. **Pustulosis palmaris et plantaris**
4721. **Rupioid Psoriasis**
4722. **Sebopsoriasis**
4723. **Verrucous psoriasis**
4724. **Von zumbusch pattern of generalized Pustular Psoriasis**
4725. **Psema (entity with clinical features of both psoriasis and eczema)**
4726. **PSTPIP1-associated myeloid-related-proteinemia inflammatory syndrome (PAMI Syndrome)**
4727. **Psychogenic pruritus**

4728. **Psychogenic purpura =gardner-diamond syndrome=autoerythrocyte sensitization = painful bruising syndrome**
4729. **Pten hamartoma tumor syndrome (PHTS)**
4730. **Linear PTEN nevus (linear cowden nevus included)**
4731. **Pterygium unguis**
4732. **Puffy hand syndrome**
4733. **Puffy foot syndrome**
4734. **Pulicosis= Puliciasis**
4735. **Puretic syndrome**
4736. **Purpura**
4737. **Contact purpura**
4738. **Purpura cyclops**
4739. **Purpura en cocarde**
4740. **Purpura factitia**
4741. **Purpura fulminans**
4742. **Purpura gogglorum**
4743. **Purpura progressiva pigmentosa**
4744. **Purpura pulicosa**
4745. **Purpura simplex**
4746. **Solar purpura=Solar capillaritis**
4747. **Purpura telangiectatica arciformis**
4748. **Pinch purpura**
4749. **Pustulotic arthro-osteitis (PAO)**
4750. **Puva (psoralen and UVA) related diseases**
4751. **Puva-induced acrobullous dermatosis**
4752. **Puva keratosis**
4753. **Puva induced lentigines**
4754. **pyemotes ventricosus dermatitis**
4755. **Pyknodysostosis**
4756. **Pyoderma faciale**
4757. **pyoderma fistulan=fox den disease**
4758. **Pyoderma, folliculitis and atopy, with defective leukocyte and lymphocyte function**

4759. **Pyoderma gangrenosum (see next)**
4760. ✓ **Ulcerative**
4761. ✓ **Vesiculobullous**
4762. ✓ **Pustular**
4763. ✓ **Superficial granulomatous**
4764. ✓ **Pyostomatitis vegetans**
4765. ✓ **Pyodermatitis-pyostomatitis vegetans**
4766. **Pyogenic granuloma (lobular capillary haemangioma = lobular capillary hemangioma =Botryomycoma= granuloma telangiectaticum= Granuloma pyogenicum**
4767. **Warner and wilson-jones syndrome (recurrent pyogenic granuloma with satellite lesions)**
4768. **Pyogenic granuloma-like kaposi sarcoma (PG-like ks)**
4769. **Pyogenic granuloma ike metastases**
4770. **Pregnancy-Related Pyogenic Granuloma (Granuloma Gravidarum)**
4771. **Pyogenic infections**
4772. **Pyomyositis**
4773. **Pyramidal protrusion=Perineal skin tag**
4774. **Pyrin-associated autoinflammation with neutrophilic dermatosis**
4775. **Pyruvoyltetrahydropterin synthase (6-ptps) deficiency**
4776. **Pythiosis**
4777. **Q fever**
4778. **Quincke sign**
4779. **Rabson -Mendenhall syndrome**
4780. **Rac2 deficiency**
4781. **RACAND (Raynaud's phenomenon, anticentromere antibodies,digital necrosis,without digital sclerosis)**
4782. **Racoon eyes**
4783. **Radiation dermatitis (Acute&Chronic)**
4784. **Radiotherapy-induced oral mucositis**
4785. **Radiation recall dermatitis**
4786. **Radiation-induced comedones**

4787. **Radiation-Induced Atypical Vascular Lesion**
4788. **Fluoroscopy-induced radiation dermatitis**
4789. **Ranula**
4790. **Rat-bite fever=erythema arthriticum epidemicum=Sodoku=Haverhill fever**
4791. **Ramsay–hunt syndrome**
4792. **RASopathies**
4793. **Rautenstrauch syndrome**
4794. **Raynaud phenomenon**
4795. **Raynaud syndrome**
4796. **Reactive angioendotheliomatosis**
4797. **Malignant angioendotheliomatosis**
4798. **Reactive arthritis**
4799. **Reactive infectious mucocutaneous eruption**
4800. **Reactive perforating collagenosis. RPC**
4801. **Recalcitrant, erythematous, desquamating (RED) disorder**
4802. **Recurrent cutaneous eosinophilic vasculitis**
4803. **Recurrent Neutrophilic Dermatosis of the Face**
4804. **Recurrent respiratory papillomatosis**
4805. **Recurrent toxin-mediated perineal erythema**
4806. **Recurring digital fibrous tumors of childhood (benign juvenile digital fibromatosis)**
4807. **Red baby syndrome**
4808. **Red ear syndrome**
4809. **Red finger syndrome**
4810. **Red man syndrome**
4811. **Red neck syndrome**
4812. **Red scrotum syndrome**
4813. **Reed syndrome (Hereditary Leiomyomatosis and Renal Cell Cancer Syndrome)**
4814. **Reiter syndrome=Circinate balanitis=keratoderma blennorrhagicum= reactive arthritis=circinate ulcerative vulvitis**
4815. **Relapsing eosinophilic perimyositis**
4816. **Relapsing linear acantholytic dermatosis**

4817. **Relapsing polychondritis (RPC)**
4818. **Remitting seronegative symmetrical synovitis with pitting edema (RS3PE) syndrome**
4819. **Restrictive dermopathy=tight skin contracture syndrome**
4820. **Reticular dysgenesis**
4821. **Reticular/reticulates telangiectatic erythema**
4822. **Reticulate acropigmentation of kitamura**
4823. **Reticulate hyperpigmented scleroderma RHS**
4824. **Reticulated acanthoma with sebaceous differentiation**
4825. **Reticulated acropigmentation of Dohi**
4826. **Reticulate Genital Melanosis with Localized Depigmentation**
4827. **Reticulate vascular nevus =cutis marmorata telangiectatica congenita, congenital livedo reticularis**
4828. **Retiform hemangioendothelioma**
4829. **Restless legs syndrome**
4830. **Revesz syndrome**
4831. **Reye syndrome**
4832. **Malignant rhabdoid tumor**
4833. **Rhabdomyoma**
4834. **Rhabdomyomatous mesenchymal hamartoma=striated muscle Hamartoma**
4835. **Rhabdomyosarcoma**
4836. **Botryoid embryonal rhabdomyosarcoma**
4837. **Rheumatic fever nodule**
4838. **Rheumatoid arthritis &rheumatoid nodule & bywatery lesions & felty syndrome**
4839. **Rheumatic neutrophilic dermatosis RND**
4840. **Rhinophyma**
4841. **Rhinoscleroma**
4842. **Rhinosporidiosis**
4843. **Rhizopus fungal infection**
4844. **Rhytids**
4845. **Riboflavin deficiency= Vitamin B2 deficiency**
4846. **Richter syndrome**

4847. **Rickettsia aeschlimannii infection**
4848. **Rickettsia parkeri rickettsiosis**
4849. **Rickettsia massiliae rickettsiosis**
4850. **Rickettsial pox**
4851. **Rickettsia species 364D rickettsiosis**
4852. **Riehl's melanosis = pigmented contact dermatitis**
4853. **Riga fede disease**
4854. **Robert syndrome with tetraphacomelia= pseudothalidomide syndrome**
4855. **Rocky mountain spotted fever**
4856. **Rogue hairs**
4857. **Rombo syndrome**
4858. **Rosacea (erythematotelangiectatic & papulopustular & phymatous & ocular & granulomatous)**
4859. **Rosacea fulminans=pyoderma faciale**
4860. **Rosacea like lupus**
4861. **Rosacea like tinea facei**
4862. **Rosacea-like demodicidosis of the scalp**
4863. **Rosacea conglobate**
4864. **Halogen rosacea**
4865. **Gram negative rosacea**
4866. **Rosenthal-kloepfer syndrome**
4867. **Rosselli–gulienetti syndrome**
4868. **Roseola infantum= exanthem subitum= 6th disease sixth= Pseudorubella**
4869. **Roseola vaccinatum**
4870. **Ross River virus infection**
4871. **Ross syndrome**
4872. **Rose handler's disease**
4873. **Rothmann-makai syndrome**
4874. **Rothmund-Thomson syndrome =poikiloderma congenitale**
4875. **Roth-bernhardt syndrome**
4876. **Rowell syndrome**
4877. **Rubella= German measles=three-day measles=Third Disease**
4878. **Rubinstein–taybi syndrome=broad thumb–hallux syndrome**

4879. **Ruocco's immunocompromised cutaneous district**
4880. **Rud syndrome**
4881. **Rüdiger syndrome**
4882. **Rudimentary toe**
4883. **Ruggieri-happle syndrome**
4884. **Rumpel leede sign**
4885. **Russell–silver syndrome**
4886. **Russel sign**
4887. **Sabinas brittle hair and mental deficiency syndrome**
4888. **Salivary gland neoplasms (pleomorphic adenoma&canalicular adenoma&mucoepidermoid carcinoma&polymorphous adenocarcinoma&adenoid cystic carcinoma)**
4889. **Salt and pepper leukoderma**
4890. **Saddan syndrome**
4891. **Saddle nose**
4892. **Saethre-chotzen syndrome**
4893. **Sago palm disease**
4894. **SAHA syndrome :seborrhea, acne, hirsutism, and alopecia**
4895. **Sakati syndrome**
4896. **Salamon syndrome**
4897. **SAM syndrome (severe dermatitis–multiple allergies–metabolic wasting)**
4898. **Sandmann–Andra syndrome**
4899. **Sandfly fever =phlebotomus fever**
4900. **Sarcoidosis (see next)**
4901. **✓Papules**
4902. **✓ plaques**
4903. **✓ lupus pernio**
4904. **✓ Scar**
4905. **✓ Tattoo**
4906. **✓Lichenoid**
4907. **✓Alopecic**
4908. **✓ Annular**

4909. ✓ Atrophic
4910. ✓ lichenoid
4911. ✓ Psoriasiform
4912. ✓ Subcutaneous (darier-roussy)
4913. ✓ Angiolupoid
4914. ✓ Erythrodermic
4915. ✓ Hypopigmented
4916. ✓ Ichthyosiform
4917. ✓ Micropapular
4918. ✓ Ulcerative
4919. ✓ Verrucous
4920. ✓ Morpheaform
4921. ✓ Sarcoidois early onset type
4922. ✓ Pseudotumoral cutaneous sarcoidosis
4923. Sarcoma
4924. Sarcomata
4925. Scabies **(see next)**
4926. ✓ Crustd / Norwegian scabies
4927. ✓ Bullous scabies
4928. ✓ Infantile scabies
4929. ✓ Scabies inclean
4930. ✓ Scabetic nodule
4931. ✓ Pseudo scabies
4932. ✓ Pyoscabies=infected scabies
4933. ✓ Scabies surrepticius
4934. ✓ Scabies herpeticum
4935. ✓ Scabies incognito
4936. ✓ Scabid
4937. Saurian papulosis
4938. Scalp–ear–nipple syndrome (finlay–marks syndrome)
4939. Scarlet fever=Scarlatina

4940. **SCALP syndrome (sebaceous nevus-cns malformations-aplasia cutis congenital-limbal dermoid-pigmented nevus)**
4941. **Scalp dysesthesia**
4942. **Hypertrophic scar**
4943. **Atrophic scar**
4944. **Simple scar**
4945. **Scars of prematurity**
4946. **SCARF syndrome(skeletal abnormalities, cutis laxa, craniostenosis ,ambiguous genitalia, retardation, facial abnormalities)**
4947. **Scedosporiosis**
4948. **Schimke immuno-osseous dysplasia (**
4949. **Schimmelpenning–feuerstein–mims syndrome SFM**
4950. **Schindler disease (α -Nacetylgalactosaminidase =α - NAGA deficiency)**
4951. **Schinzel–giedion midface retraction syndrome**
4952. **Schnitzler's syndrome**
4953. **Schöpf–schulz–passarge syndrome**
4954. **Schwannoma = neurilemmoma**
4955. **Neuroblastoma-Like Schwannoma**
4956. **Plexiform Schwannoma**
4957. **Hybrid schwannoma /perineurioma**
4958. **Nodular scleroderma**
4959. **Scleredema**
4960. **Scleredema of Bushke**
4961. **Sclerema neonatorum**
4962. **Scleroderma**
4963. **Scleroderma-like cutaneous metastasis**
4964. **Sclerodermatomyositis**
4965. **Sclerodermoid disorders**
4966. **Scleromalacia**
4967. **Scleromyositis**
4968. **Scleromyxedema**
4969. **Sclerosing epithelioid fibrosarcoma**
4970. **Sclerosing lipogranuloma**

4971. **Systemic Sclerosis**
4972. **Diffuse Systemic sclerosis**
4973. **Limited Systemic sclerosis**
4974. **Systemic Sclerosis Overlap Syndromes**
4975. **Intermediate cutaneous scleroderma (icSSc)**
4976. **Systemic Sclerosis Sine scleroderma SSc (ssSSc)**
4977. **Sclerotic fibroma=Storiform Collagenoma**
4978. **Scrotal cyst /idiopathic scrotal calcification**
4979. **Scrotal fat necrosis**
4980. **Scrub typhus= Tsutsugamushi disease**
4981. **Scurvy**
4982. **Scytalidium infections**
4983. **Seaweed dermatitis**
4984. **Sea urchin granuloma**
4985. **Sealpox**
4986. **Sebastian syndrome**
4987. **Sebaceous filaments**
4988. **Hyperactive Sebaceous glands of newborn**
4989. **Sebaceoma**
4990. **Sebaceous carcinoma**
4991. **Sebaceous epithelioma**
4992. **Sebaceous hyperplasia**
4993. **Sebaceous gland prominence**
4994. **Sebaceous hyperplasia variant: juxtaclavicular beaded lines= Striated beaded lines**
4995. **Sebacous adenoma**
4996. **Seboacanthoma**
4997. **Sebocystomatosis**
4998. **Sebofolliculoma**
4999. **Sebomatricoma**
5000. **Seborrheic dermatitis =Seborrhoeic eczema= seborrhea**
5001. **Petaloid seborrheic dermatitis**
5002. **Seborrhoea Herpeticum**

5003. **Seborrheic keratosis SK (Pedunclated & Pigmented & Superficial & Eruptive & Irritated&Tadpoleseborrheic keratosis)**
5004. **SASKIA nevus (segmentally arranged seborrheic keratoses with impending atypia**
5005. **Seabather's Eruption=sea lice**
5006. **Seborrheic melanosis**
5007. **Seckel syndrome**
5008. **Secretan syndrome**
5009. **Secretory carcinoma of the skin**
5010. **Segmental congenital vascular anomaly with atrophy, ulceration and scarring (SeCVAUS)**
5011. **Segmental Pigmentation Disorder**
5012. **Selective IGA deficiency**
5013. **Selective IGM deficiency**
5014. **Selenium toxicity/excess**
5015. **Selenium Deficiency= Keshan disease=Kashin–Beck disease**
5016. **Sener syndrome (frontonasal dysplasia and dilated virchow –robin spaces)**
5017. **Senile gluteal dermatosis**
5018. **Senile purpura**
5019. **Sennetsu fever**
5020. **Sensitive skin syndrome**
5021. **Sensorineural hearing loss, enamel hypoplasia and nail defects (heimler syndrome)**
5022. **Septic Vasculopathy**
5023. **Seroma= lymphocoele**
5024. **Serpentine supravenous hyperpigmentation**
5025. **Serratia marcescens**
5026. **Servelle martorell syndrome=Port-wine stain, venous malformations and limb hypotrophy**
5027. **Setleis' syndrome**

5028. **Severe combined immunodeficiency SCID (X-linked SCID &JAK3 deficiency&Interleukin-7 receptor deficiency &ADA deficiency & PNP deficiency&Reticular dysgenesis &Coronin 1A deficiency & Omenn syndrome&Artemis deficiency&PRKDC deficiency&LIG4 syndrome&NHEJ1 syndrome&TCR deficiencies &CD45 deficiency&ZAP-70 deficiency& CD8 deficiency&Calcium channel deficiency&MHC class I deficiency &MHC class II deficiency&Winged helix nude deficiency)**

5029. **Severe dermatitis, multiple allergies, and metabolic wasting (SAM)**
5030. **Sexual abuse**
5031. **Sexual Transmitted diseases STDs**
5032. **Shaheen syndrome**
5033. **Shitake dermatitis**
5034. **Short anagen syndrome**
5035. **Short stature with pleonosteosis and periarticular fibrosis = léri's syndrome**
5036. **Shprintzen–Goldberg syndrome**
5037. **Shwachman-diamond syndrome**
5038. **Sialidosis**
5039. **Sialolith**
5040. **Sideroblastic anemia with B cell immunodeficiency, periodic fevers and developmental delay**
5041. **Siderosis**
5042. **Silica granuloma**
5043. **Silicone granuloma= siliconoma**
5044. **Silver nirate pigmentation**
5045. **Simpson-golabi-behmel syndrome**
5046. **Sindbis virus infection**
5047. **Singleton-merten syndrome (SMS)**

"sines" in dermatology (see next)

5048. ✓ **Acne fulminans sine fulminans**
5049. ✓ **Dermatomyositis sine myositis**
5050. ✓ **Eccrine hidradenitis sine neutrophils**

5051. ✓ **Eruptive seborrheic keratosis sine malignancy**
5052. ✓ **linear melorheostotic scleroderma with hypertrichosissine melorheostosis**
5053. ✓ **Lupus sine lupo**
5054. ✓ **Malignant acanthosis nigricans sine malignancy**
5055. ✓ **Necrobiosis lipoidica sine diabetes**
5056. ✓ **Pellagra sine pellagra**
5057. ✓ **Pruritus sine materia**
5058. ✓ **Psoriatic arthritis sine psoriasis**
5059. ✓ **Systemic sclerosis sine scleroderma**
5060. ✓ **Zoster sine herpete**
5061. **Sinusoidal hemangioma**
5062. **Sinus pericranii**
5063. **Sipple's syndrome**
5064. **Sirenomelia (mermaid syndrome)**
5065. **Sister marry joseph nodule**
5066. **Sitosterolemia**
5067. **Sjogren syndrome**
5068. **Skin-ache syndrome**
5069. **Skin fragility syndrome**
5070. **SKINTED (Surgery of the Knee, Injury to the Infrapatellar branch of the saphenous Nerve, Traumatic Eczematous Dermatitis)**
5071. **Slapped cheek syndrome**
5072. **Slapped neck solar melanosis**
5073. **Smith–lemli–opitz syndrome**
5074. **Small pox infection=Variola major**
5075. **Smegma pearls**
5076. **Smooth muscle hamartoma=Congenital arrector pili Hamartoma= Muscle naevi**
5077. **Sneddon wilkinson disease**
5078. **Sneddon syndrome= Idiopathic livedo reticularis with systemic involvement**

5079. **Sock line bands**
5080. **Sock line hyperpigmentation**
5081. **SOFT syndrome (short stature, onychodysplasia, facial dysmorphism, and hypotrichosis)**
5082. **Solar /Actinic elastosis**
5083. **Solar elastotic bands**
5084. **Solamen syndrome**
5085. **Solitary fibrous tumor**
5086. **Sotos syndrome**
5087. **Sparganosis**
5088. **Specific granule deficiency**
5089. **Spider angioma= spider nevus= nevus araneus=spider telangiectasia**
5090. **Spiegler–fendt sarcoid -lymphocytoma cutis**
5091. **Spinal dysraphism**
5092. **Spindle cell hemangioma**
5093. **Spindle hemangioendothelioma**
5094. **Spindle cell lipoma**
5095. **Spiradenoma**
5096. **Spiradenocylindroma**
5097. **Spiradenocarcinoma**
5098. **Malignant eccrine spiradenoma (MES)**
5099. **Split hand -foot malformation (SHFM1)**
5100. **Spondyloenchodrodysplasia with immune dysregulation**
5101. **Spontaneous scrotal ulceration**
5102. **Sporobolomyces salmonicolor**

Spots in dermatology (see next)

5103. ✓ **Nickel spot test**
5104. ✓ **Cotton wool spots**
5105. ✓ **Stasis spots**
5106. ✓ **Antimony spots**
5107. ✓ **Ash leaf spots**
5108. ✓ **Bier spots**

5109. ✓Bitot's spots
5110. ✓Black spot test
5111. ✓Black-spot poison ivy
5112. ✓Brushfield spots
5113. ✓Café au lait spots
5114. ✓Café noir spot
5115. ✓Campbell de morgan spots
5116. ✓Cayenne pepper spots
5117. ✓Drusen-like spots
5118. ✓Forchheimer spots
5119. ✓Fordyce spots
5120. ✓Herman spots
5121. ✓Ink spot lentigo=sunburn lentigo
5122. ✓Janeway spots
5123. ✓Koplik spots
5124. ✓Leopard skin spotting
5125. ✓Liver spots
5126. ✓Lucio's spotted leprosy
5127. ✓Milk spots
5128. ✓Mongolian spot
5129. ✓Monospot test
5130. ✓Nagayama spots
5131. ✓Oil spots
5132. ✓Orange palpebral spots
5133. ✓Rose spots
5134. ✓Roseolar spots
5135. ✓Roth spots
5136. ✓Shin spots (binkley's spots, pigmented pretibial patches, spotted leg syndrome, diabetic dermangiopathy)
5137. ✓Sporty spots
5138. ✓Spot urine tests

5139. ✓Spot tests in allergic contact dermatitis
5140. ✓Spotted fevers
5141. ✓Spotted lunula
5142. ✓Spotty parakeratosis=staccato parakeratosis:
5143. ✓Twin spots (syn: didymosis)
5144. ✓White spot disease (syn: lichen sclerosus)
5145. Sporotrichosis
5146. Squamous cell papilloma
5147. Squamo-melanocytic tumor
5148. Squamous metaplasia of the lactiferous ducts
5149. Squamous cell carcinoma
5150. Verrocous squamous cell carcinoma
5151. Squamoid eccrine ductal carcinoma
5152. Staphylococal scalded skin syndrome SSSS=Ritter disease=pemphigus neonatorum
5153. Stasis purpura
5154. Stasis dermatitis
5155. Steal syndrome
5156. Steatocystoma (simplex & multiplex & multiplex suppurativum)
5157. Sternal cleft
5158. Stellate scar
5159. Steriod induced rosacea
5160. Stevens-johnson syndrome SJS
5161. SJS /TEN overlap
5162. Fuchs‘ syndrome (stevens-johnson syndrome without skin involvement)
5163. Stewart-bluefarb syndrome
5164. Stewart-treves syndrome=Postmastectomy lymphangiosarcoma
5165. Stickler syndrome
5166. Stiff baby syndrome
5167. Stiff skin syndrome =congenital fascial dystrophy
5168. Adult-onset Still disease
5169. Systemic-onset juvenile idiopathic arthritis (sJIA)= Still disease

5170. **Sting-associated vasculopathy with onset in infancy SAVI**
5171. **STK4 deficiency**
5172. **Stomal related diseases**
5173. **Stomatitis medicamentosa**
5174. **Gold-induced stomatitis**
5175. **Allergic Contact Stomatitis**
5176. **Antibiotic-induced stomatitis**
5177. **Storiform collagenoma**
5178. **Storm's syndrome**
5179. **Straight hair nevus**
5180. **Strawberry in dermatology (see next)**
5181. ✓ **Strawberry gingival hyperplasia**
5182. ✓ **Strawberry cervix**
5183. ✓ **Strawberry nevus**
5184. ✓ **Strawberry papillomatosis**
5185. ✓ **Strawberry like nasal growth**
5186. ✓ **Strawberry tongue**
5187. **Streptocerciasis**
5188. **Streptococcal sex syndrome**
5189. **Striae = strech mark= Atrophoderma maculatum**
5190. ✓**Striae alba**
5191. ✓**Striae rubra**
5192. ✓**Striae distensae**
5193. ✓**Striae gravidarum**
5194. **Strongyloidiasis**
5195. **Stucco keratosis**
5196. **Sturge weber syndrome (encephalotrigeminal angiomatosis)**
5197. **Subaponeurotic (subgaleal) fluid collection**
5198. **Subcorneal pustular dermatosis= sneddon–wilkinson disease**
5199. **Subcutaneous emphysema**
5200. **Subcutaneous fat necrosis of the newborn**
5201. **Subepidermal calcified nodule**

5202. **Subgaleal empyema**
5203. **Subgaleal hematomas**
5204. **Sucking blisters**
5205. **Suction purpura/factitious purpura**
5206. **Sudden whitening of hair**
5207. **Sudoriparous angiomas SA**
5208. **Sulzberger–garbe syndrome= Exudative discoid and lichenoid chronic dermatosis= oid-oid disease**
5209. **Sunburn= Dermatitis solaris**
5210. **Superficial CD34-positive fibroblastic tumor**
5211. **Superficial migratory thrombophlebitis**
5212. **Superficial papular neuroma**
5213. **Superior vena cava syndrome**
5214. **Supernumeray nostril**
5215. **Supravenous lichen planus**
5216. **Sweat: (hyperhidrosis&hypohidrosis&anhidrosis)**
5217. **Sweet syndrome :Acute Febrile neutrophilic dermatosis**
5218. **Swimmer's ear**
5219. **Swimmer's itch =cercarial dermatitis= Dermatitis schistosomica**
5220. **Swimmers's xerosis**
5221. **SycosisBarbae=Sycosis Vulgaris= Barber's Itch**
5222. **Mycotic Sycosis**
5223. **Herpetic Sycosis**
5224. **Lupoid Sycosis**
5225. **Symmetric acral annular erythema**
5226. **Symmetric idiopathic micropapular facial eruption (SIMPLE)**
5227. **Symmetrical acrokeratoderma (SAK)**
5228. **Symmetrical palatal fibromatosis**
5229. **Symmetrical peripheral gangrene**
5230. **Symmetrical progressive leukopathy**
5231. **Symmetric synovitis**
5232. **Symplastic hemangioma**
5233. **Synovial cyst**

5234. **Synovial metaplasia**
5235. **Synovial sarcoma**
5236. **Synovitis, acne, pustulosis, hyperostosis and osteitis SAPHO syndrome**
5237. **Syphilis = great pox (primary chancre& secondary & &congenital& lues maligna)**
5238. **Secondary syphilis : (collarette of biett & condylomata lata & corona veneris & necklace of venus & syphilitic perlèche & plaques muqueuses)**
5239. **Tertiary syphilis (late): gummas**
5240. **Syphilitic balanitis of follmann (SBF)**
5241. **Malignant syphilis=Lues maligna of syphilis**
5242. **Congenital syphilis (rhagades & hutchinson teeth & mulberry molars & saddle nose &olympian brow& higouménakis' sign &saber shins & clutton's joints)**
5243. **Herpetic syringitis**
5244. **Syringoacanthoma of rahbari**
5245. **Syringocystadenoma papilliferum SCAP**
5246. **Syringocystadenocarcinoma papilliferum SCAP**
5247. **Syringoma**
5248. **Eruptive syringomas =Eruptive hidradenoma of Darier and Jaquet**
5249. **Eruptive milium-like syringoma (EMS)**
5250. **Infiltrating Syringomatous Adenoma of Nipple**
5251. **Syringofibroadenoma= acrosyringeal nevus of weedon and lewis**
5252. **Syringoid eccrine carcinoma= eccrine carcinoma**
5253. **Syringomatous carcinoma**
5254. **syringolymphoid Hyperplasia with alopecia and anhidrosis**
5255. **Syringomyelia**
5256. **Skin tag = acrochordon = fibroepithelial polyp =soft fibroma=Lax Fibroids**
5257. **Takayasu arteritis**
5258. **Tangier disease =alpha-Lipoprotein Deficiency Disease**
5259. **Tail /faun tail**
5260. **Talaromyces marneffei infection**
5261. **Talon noir**
5262. **Tanapox**

5263. **Tapered fracture hair**
5264. **TAR syndrome (Thrombocytopenia with absent radius syndrome)**
5265. **Tattoo**
5266. **Taurodontia, absent teeth and sparse hair**
5267. **Tay syndrome**
5268. **Teebi–Shaltout syndrome**
5269. **Telangiectasia**
5270. **Eruptive recurrent pigmented telangiectasia**
5271. **Telangiectasia macularis eruptive perstan TMEP**
5272. **Telangiectasia macularis multiplex acquisita**
5273. **Telogen effluvium**
5274. **TEMPI syndrome (Telangiectasias, Erythrocytosis , Monoclonal gammopathy, Perinephric-fluid collections , and Intrapulmonary shunting)**
5275. **Tennis toe=Runner's toe=jogger's toe**
5276. **Teratoma**
5277. **Terminal osseous dysplasia with pigmentary defects TODPD**
5278. **Terra firma forme dermatosis**
5279. **Tetramelic deficiencies, ectodermal dysplasia, deformed ears and other abnormalities (odontotrichomelic syndrome)**
5280. **Texier disease**
5281. **Thanos syndrome**
5282. **Thinker sign**
5283. **Thromboangitis obliterans**
5284. **Thrombocytopenic purpura**
5285. **Thrombotic thrombocytopenic purpura**
5286. **ICI-(Immune Checkpoint Inhibitor)induced thrombotic vasculopathy**
5287. **Thumb- and finger sucking**
5288. **Thumb deformity-alopecia-pigmentation anomaly syndrome**
5289. **Thymic dysplasia**
5290. **Thymic cyst**
5291. **Thyroglossal cyst**
5292. **Tibialis anterior muscle herniation**
5293. **Tick borne lymphadenopathy (TIBOLA)**

5294. Tick-borne relapsing fever
5295. Tietz syndrome
5296. Tinea amiantacea =pityriasis amiantacea
5297. Tinea barbae
5298. Tinea blepharociliaris
5299. Tinea capitis (scaly & black dot& grey patch&pustular& kerion& agminate folliculitis)
5300. Tinea favosa= favus
5301. Tinea corporis= ringworm of the body
5302. Tinea corporis bullosa
5303. Tinea corporis hyperkeratotica
5304. Tinea cruris= ringworm of the groin
5305. Tinea gladiatorum
5306. Tinea faciei = ringworm of the face
5307. Tinea incognito
5308. Tinea imbricata
5309. Tinea pseudoimbricata
5310. Majocchi granuloma
5311. Tinea manuum
5312. Tinea nigra= Keratomycosis mgncans palmaris
5313. Tinea pedis= athlete's foot (moccasin & interdigital & vesicular & ulcerative & purpuric)
5314. One-hand-two-feet tinea syndrome
5315. Tinea profunda
5316. Tinea versicolor TVC =pityriasis versicolor PVC=dermatomycosis furfuracea= tinea flava=tinea alba =achromia parasitica
5317. ATrophic tinea versicolor TVC
5318. Erythrasmoid pityriasis versicolor
5319. Follicular Tinea versicolor TVC/Confetti-like pityriasis versicolor
5320. Tinea unguium
5321. Tumor necrosis factor receptor-associated periodic fever syndrome (TRAPS)= familiar Hibernian fever
5322. Tongue: annulus migran

5323. **Bifid tongue**
5324. **Black hairy tongue = lingua villosa nigra**
5325. **Caviar tongue=sublingual varicosities**
5326. **Scrotal Tongue: fissured= plicated tongue=furrowed**
5327. **Furred tongue**
5328. **Crenated tongue**
5329. **Fungiform papilla of tongue**
5330. **Geographic Tongue (stomatitis areata migrans=benign migratory glossitis)**
5331. **Smooth Tongue**
5332. **Topical steroid damaged face (TSDF)**
5333. **Toponymous contributions (see next)**
5334. ✓ **Acne mallorca**
5335. ✓ **Acroangiodermatitis of Mali**
5336. ✓ **African eye worm infection**
5337. ✓ **African Endemic Kaposi sarcoma**
5338. ✓ **African histoplasmosis**
5339. ✓ **African tick bite fever**
5340. ✓ **African trypanosomiasis**
5341. ✓ **Aleppo boil**
5342. ✓ **Ambras syndrome**
5343. ✓ **American hookworm infection**
5344. ✓ **American trypanosomiasis**
5345. ✓ **Andean sickness=white leprosy**
5346. ✓ **Balsam of Peru**
5347. ✓ **Biskra button**
5348. ✓ **Brazilian purpuric fever**
5349. ✓ **Bruton disease**
5350. ✓ **Burdwan fever**
5351. ✓ **Buruli ulcer**
5352. ✓ **Calabar swelling**

5353. ✓ Calcutta ulcer
5354. ✓ Congo red stain
5355. ✓ Coast of california calms neurofibromatosis
5356. ✓ Coast of maine mccune-albright syndrome
5357. ✓ Delhi boil
5358. ✓ Dum dum fever
5359. ✓ European blastomycosis
5360. ✓ Familial mediterranean fever
5361. ✓ Fort chaffee fever
5362. ✓ French disease for syphilis
5363. ✓ Gambian trypanosomiasis
5364. ✓ Haverhill fever
5365. ✓ Hibernian fever
5366. ✓ Indian filing
5367. ✓ Indian tick typhus
5368. ✓ Jaipur block
5369. ✓ Jodhpur technique
5370. ✓ Kandahar sore
5371. ✓ Lahore sore
5372. ✓ Lyme disease
5373. ✓ Madura foot
5374. ✓ Mal de maleda ppk
5375. ✓ Marseilles fever
5376. ✓ Mongolian spots
5377. ✓ Morbihan disease
5378. ✓ Nagashima palmoplantar keratoderma
5379. ✓ Naxos disease
5380. ✓ North america blastomycosis
5381. ✓ North carolina fever
5382. ✓ Norwegian scabies
5383. ✓ Ogna type epidermolysis bullosa simplex

5384. ✓ Ohio river valley fever
5385. ✓ Peruvian warts
5386. ✓ Pahvant valley fever
5387. ✓ Rocky mountain spotted fever
5388. ✓ Rose Bengal stain
5389. ✓ San joaquin valley fever
5390. ✓ Silk route disease
5391. ✓ South American blastomycosis
5392. ✓ Tokelau
5393. ✓ Tularemia
5394. ✓ Volhynia fever
5395. ✓ West nile fever
5396. ✓ Ebola virus disease
5397. Torch syndrome
5398. Torus mandibularis
5399. Torus palatine
5400. Touraine-solente-gole syndrome
5401. Toxic epidermal necrolysis TEN
5402. Toxic oil epidemic syndrome
5403. Toxic shock syndrome
5404. Toxocariasis
5405. Toxoplasmosis
5406. Transient abdominal telangiectasia of the newborn
5407. Transient acantholytic dermatosis
5408. Transient aquagenic wrinkling of palms
5409. Transient bullous dermolysis of the newborn
5410. Transient hypogammaglobulinemia of infancy
5411. Transient infantile lingual leukoplakia
5412. Transient lingual papillitis
5413. Transient localized hyperpigmentation newborn
5414. Transient neonatal cutis l axa
5415. Transient neonatal hair loss

5416. **Transient neonatal porphyrinaemia**
5417. **Transient neonatal pustular melanosis=Lentigines neonatorum**
5418. **Transient neonatal zinc deficiency**
5419. **Transient pigmentary lines of the newborn**
5420. **Transient telangiectatic purpura**
5421. **Transitory contractile dermatoses**
5422. **Transplantation related diseases**
5423. **Transporter associated with antigen processing TAP deficiency**
5424. **Traumatic ulcer**
5425. **Traumatic ulcerative granuloma**
5426. **Treacher Collin syndrome**
5427. **Tree man syndrome**
5428. **Trench fever**
5429. **Trench foot**
5430. **Manitoba oculo tricho anal syndrome**
5431. **Trichiasis**
5432. **Trichinosis**
5433. **Tricho entero hepatic syndrome**
5434. **Tricho oculo dermo vertebral syndrome**
5435. **Tricho retino dento digital syndrome**
5436. **Trichoadenoma**
5437. **Verrucous trichoadenoma**
5438. **Trichoadenoma of nikolowski**
5439. **Trichobacteriosis pubis**
5440. **Trichoblastic carcinoma**
5441. **Trichoblastic carcinosarcoma**
5442. **Trichoblastic infundibular cyst TBIC**
5443. **Trichoblastoma**
5444. **Desmoplastic trichoblastoma**
5445. **Trichoclasis**
5446. **Trichodental syndrome**
5447. **Tricho-dento-osseous syndrome**
5448. **Trichodiscoma**

5449. **Trichodynia**
5450. **Trichodysplasia spinulosa**
5451. **Trichodysplasia‑amelogenesis imperfecta**
5452. **Trichoepithlioma**
5453. **Desmoplastic trichoepithelioma=sclerosing epithelial hamartoma**
5454. **Trichofolliculoma**
5455. **Trichogerminoma**
5456. **Tricholemmoma**
5457. **Desmoplastic tricholemmoma**
5458. **Malignant proliferating trichilemmal tumor**
5459. **Trichilemmal carcinoma**
5460. **Trichilemmal cyst**
5461. **Trichomadesis**
5462. **Trichomalacia**
5463. **Trichomatricoma**
5464. **Trichomegaly**
5465. **Trichomoniasis**
5466. **Trichomycosis axillaris**
5467. **Trichonodosis**
5468. **Tricho-odonto-onychial dysplasia**
5469. **Tricho-odonto-onycho dermal syndrome**
5470. **Tricho-onycho dental syndrome**
5471. **Trichopathophobia**
5472. **Trichopathy**
5473. **Trichophagia**
5474. **Trichophyton verrucosum**
5475. **Trichoptilosis**
5476. **Trichorrhexis invaginata**
5477. **Trichorrhexis nodosa**
5478. **Trichorhinophalangeal syndrome= langer–giedion**
5479. **Trichorhizophagia**
5480. **Trichorrhea**
5481. **Trichoschisis**

5482. **Trichoschisis/onychodystrophy**
5483. **Trichosporonosis**
5484. **Trichostasis spinulosa**
5485. **Trichothiodystrophy**
5486. **Trichotillomania (see next)= Alopecia Artefacta**
5487. ✓ **Trichoteiromania**
5488. ✓ **Trichotemnomania**
5489. ✓ **Trichodaganomania**
5490. ✓ **Telogen mania**
5491. ✓ **Trichophobia**
5492. ✓ **Trichorrexomania**
5493. ✓ **Trichotillomania Incognito**
5494. ✓ **Trichobezoar**
5495. ✓ **Trichophytobezoar**
5496. **Trisomy 13 (patau syndrome)**
5497. **Tripe palms**
5498. **Trigeminal neuralgia**
5499. **Trigeminal trophic syndrome**
5500. **Trimethylaminuria**
5501. **Trousseau's sign**
5502. **Traumatic ulcerative granuloma with stromal eosinophilia**
5503. **Trumpeter's wart**
5504. **African Trypanosomiasis = Sleeping sickness**
5505. **American Trypanosomiasis= Chagas disease**
5506. **Tuberous sclerosis complex=(bourneville disease & epiloia & angiofibroma & shagreen patch & ash-leaf macules &koenen tumour & periungual fibroma & molluscum pendulum & fibrous cephalic plaques & white freckles&Folliculocystic and collagenous hamartoma FCCH)**
5507. **Tuberculosis TB (see next)**
5508. ✓ **Gumma**
5509. ✓ **lupus vulgaris=tuberculosis lupus (Plaque& Hypertrophic& Ulcerative&Vegetative)**

5510. ✓ Acute military cutaneous tuberculosis
5511. ✓ Tuberculosis cutis orificialis
5512. ✓ Papulonecrotic tuberclid
5513. ✓Rosacea-like tuberculid of Lewandowsky
5514. ✓ Lichen scrofulosorum=Tuberculosis cutis lichenoides
5515. ✓ Primary cutaneous tuberculosis complex=chancre
5516. ✓ Scrofuloderma=tuberculosis colliquativa cutis
5517. ✓ TB verrucous cutis=warty lupus
5518. ✓ Bcgitis –BCG
5519. ✓Tuberculous pseudomycetoma
5520. ✓Tuberculous mastitis
5521. Mycobacterial spindle cell pseudotumor
5522. Mycobacterium abcess
5523. Mycobacterium avium -intracellulare complex
5524. Mycobacterium chelonae
5525. Mycobacterium fortuitum
5526. Mycobacterium gordonae disease
5527. Mycobacterium haemophilum
5528. Mycobacterium indicus pranii
5529. Mycobacterium kansasii
5530. Mycobacterium llatzerense
5531. Mycobacterium malmoense disease
5532. Mycobacterium marinum =fish tank granuloma=Swimming pool granuloma
5533. Mycobacterium scrofulaceum
5534. Mycobacterium szulgai disease
5535. Mycobacterium ulceran= buruli ulcer
5536. Mycobacterium xenopi disease
5537. Tubular carcinoma
5538. Tufted angioma =hypertrophic hemangioma=progressive capillary hemangioma=angioblastoma of Nakagawa
5539. Tularemia

5540. **Tufted folliculitis**
5541. **Tumour of the follicular infundibulum=infundibuloma; isthmicoma**
5542. **Tumoral melanosis**
5543. **Tungiasis**
5544. **Tunneling hair syndrome**
5545. **Turban tumor**
5546. **Turcot syndrome**
5547. **Turkey Gobbler neck /cervicomental**
5548. **Turner syndrome**
5549. **Twisted and rolled body hairs**
5550. **Tyloma**
5551. **Tylosis**
5552. **Type 1 bare lymphocyte syndrome**
5553. **Typhoid &salmonellosis**
5554. **Epidemic (louse -borne) typhus**
5555. **Flying squirrel typhus**
5556. **Murine typhus=Endemic Typhus**
5557. **Scrub typhus= Tsutsugamushi disease= Mite-borne typhus**
5558. **North Asian Tick Typhus (Siberian Tick Typhus)**
5559. **Queensland Tick Typhus**
5560. **Sporadic typhus=Brill–Zinsser disease**
5561. **Sylvatic Typhus**
5562. **Tyrosinemia**
5563. **Tyson's Glands**
5564. **Ulcer**
5565. **Ulcer in antiphospholipid syndrome**
5566. **Arterial Ulcer**
5567. **Chronic plantar Ulcer**
5568. **Decubitus/pressure Ulcer**
5569. **Diabetic ulcer**
5570. **Genital Ulcer**
5571. **Ulcerated gouty tophi**
5572. **Ulcer due to Hematologic Disorders**

5573. **Ischemic ulcer**
5574. **Lipschutz ulcer / Lipschütz**
5575. **Marjolin ulcer**
5576. **Meleney ulcer**
5577. **Martorel ulcer**
5578. **Neuropathic ulcer**
5579. **Ulcer in rheumatoid arthritis**
5580. **Ulcer in sickel cell anemia**
5581. **Tropical Ulcer=tropical phagedaena**
5582. **Ulcer in Thalassemia**
5583. **Ulcer in vasculitis**
5584. **venous ulcer**
5585. **Ulerythema ophryogenes =keratosis pilaris atrophicans faciei)**
5586. **Ulerythema sycosiforme**
5587. **Ulnar–mammary syndrome**
5588. **Umblical granuloma**
5589. **Uncombable hair syndrome**
5590. **Uncombable hair, retinal pigmentary dystrophy, dental anomalies and brachydactyly (bork syndrome)**
5591. **Undifferentiated connective tissue disease(UCTD)**
5592. **Unilateral laterothoracic exanthema=asymmetric periflexural exanthem of childhood**
5593. **Unilateral naevoid telangiectasia**
5594. **Universal angiomatosis**
5595. **Universal Lightening of Hair**
5596. **Uncombable hair syndrome**
5597. **Unverricht–Lundborg Disease**
5598. **Upper dermal elastolysis**
5599. **Upshaw- schulman syndrome**
5600. **Urachus cyst**
5601. **Urbach-wiethe disease**
5602. **Uremic follicular hyperkeratosis**
5603. **Uremic frost**

5604. **Urticaria= Hives (acute & chronic & recurrent)**
5605. **Adrenergic urticaria**
5606. **Aquagenic urticaria**
5607. **Cholinergic urticarial**
5608. **Cold urticaria**
5609. **Contact urticaria**
5610. **Exercise-induced urticaria**
5611. **Exercise induced anaphylaxis**
5612. **Galvanic Urticaria**
5613. **Heat urticaria**
5614. **Papular urticaria**
5615. **Pressure urticarial**
5616. **Psychogenic Urticaria**
5617. **Recall urticaria**
5618. **Solar urticarial**
5619. **physical type urticaria**
5620. **Vibratory Urticaria**
5621. **Urticarial dermatitis**
5622. **Urticaria-like follicular mucinosis**
5623. **Urticaria multiforme**
5624. **Urticaria pigmentosa**
5625. **Urticarial vasculitis**
5626. **Uv-sensitive syndrome**
5627. **Vaccinia**
5628. **Generalized vaccinia**
5629. **Progressive vaccinia (vaccinia necrosum,vaccinia gangrenosum)**
5630. **Vaccination-associated morphea**
5631. **Vagabond's disease**
5632. **Valley fever**
5633. **Van allen-myhre syndrome**
5634. **Van der woude syndrome**
5635. **Van lohuizen syndrome**
5636. **Varicella& Varicella gangrenosum**

5637. **Sublingual varices**
5638. **Vascular malformations (see next)**
5639. **✓Capillary malformation**
5640. **✓Lymphatic malformation**
5641. **✓Mixed capillary-venous-lymphatic malformation**
5642. **✓Arteriovenous malformation**
5643. **✓capillary malformation-arteriovenous malformation(cm-avm)**
5644. **Vascular twin spotting (vts)**
5645. **Vasculitis: drug induced**
5646. **Vasculitis LCV leucocytoclastic**
5647. **Vasculitis: septic type**
5648. **Single organ vasculitis**
5649. **Localized chronic fibrosing vasculitis**
5650. **Vasculitis in SLE &rheumatoid arthritis**
5651. **Vasculitis: small vessel type**
5652. **Vasculitis:medium vessel type**
5653. **Vasculitis:large vessel type**
5654. **Vasculitis: hypersensitivity type**
5655. **Vein graft site dermatitis**
5656. **Vellus hair cyst**
5657. **Velocardiofacial syndrome**
5658. **Venomous Fish= Stinging fish**
5659. **Venous lake**
5660. **Venous malformation**
5661. **Cephalic venous malformation**
5662. **Verrucous venous malformation =verrucous haemangioma**
5663. **Vernix caseousa**
5664. **Verruca vulgaris (VV)**
5665. **Verrucal keratosis**
5666. **BRAF inhibitor-associated verrucous keratosis (BAVK)**
5667. **Verrucous carcinoma**
5668. **Verrucous cysts**

5669. **Verrucous hemangioma=Verrucous Venulocapillary Malformation**
5670. **Verrucous leukoplakia**
5671. **Verrucous perforating collagenoma (collagenome perforant verruciformé)**
5672. **Verrucous skin lesions in diabetic neuropathy (VSLDN)**
5673. **Verrucous xanthoma= verruciform xanthomas**
5674. **Verruga peruana (peruvian warts)=oroya fever =carrion disease= bartonellosis**
5675. **Vestibular papillomatosis**
5676. **VEXAS syndrome(Vacuoles,E1 enzyme,X-linked,autoin6ammatory,somatic)**
5677. **Vibration white finger**
5678. **Vibrio bacteria related infections**
5679. **Vici syndrome**
5680. **Viral exanthema**
5681. **Non-specific viral eruptions**
5682. **Viral hemorrhagic fevers**
5683. **Old-World Hemorrhagic Fevers (Arenaviruses :Lassa virus&Filoviruses :Marburg and Ebola& Bunyaviruses& Hantaviruses)**
5684. **New-World Hemorrhagic Fevers(Hantaviruses&Arenaviruses)**
5685. **Vitamin A deficiency= retinol**
5686. **Vitamin B deficiency(B1= Thiamine,B2= Riboflavin,B3= Niacin,B5= pantothenic acid,B6= Pyridoxine,B7 =biotin,B9= Folic acid,B12= Cobalamin)**
5687. **Vitamin C deficiency= Ascorbic acid**
5688. **Vitamin D deficiency =calcitriol**
5689. **Vitamin D-dependent rickets with alopecia**
5690. **Vitamin k deficiency= =phytonadione**
5691. **Vitiligo**
5692. **Inflammatory vitiligo**
5693. **Trichrome vitiligo**
5694. **Quadrichrome vitiligo**
5695. **Pentachrome vitiligo**
5696. **Hypochromic vitiligo (HV)**
5697. **Blue vitiligo**

5698. **Burn vitiligo**
5699. **Papulosquamous vitiligo**
5700. **Vitiligo en confetti**
5701. **vitiligo-like acquired dermal elastolysis**
5702. **Vogt–koyanagi–harada syndrome**
5703. **Volkmann's contracture**
5704. **Von hippel-lindau syndrome**
5705. **Von recklinghausen's disease**
5706. **Von zumbusch : pustular psoriasis**
5707. **VRESS :virus reactivation with eosinophilia and systemic symptoms**
5708. **Massive Vulval Edema**
5709. **Vulvar intraepithelial neoplasia VIN**
5710. **Vulvar melanosis**
5711. **Vulvitis granulomatosis**
5712. **Vulvovaginal gingival syndrome= Hewitt-pelisse**
5713. **Vulvodynia**
5714. **Walbaum –dehane–schlemmer syndrome**
5715. **Waardenburg syndrome (4types)**
5716. **Hypergammaglobulinemic purpura of waldenström**
5717. **Walking dandruff= Cheyletiellosis**
5718. **Warburg-cinotti syndrome**
5719. **Warfarin induced diseases**
5720. **Warfarin associated venous limb gangrene**
5721. **Warfarin necrosis**
5722. **Wart (common= verruca vulgaris & plane/flat= verruca plana& plantar & digitate& filiform & butcher's & genital =Condylomata Acuminata & doughnut & mirmecia & anal& mucous membrane&Punctate &Periungual and subungual& cystic)**
5723. **Warty dyskeratoma**
5724. **Warthin tumor (papillary cystadenoma lymphomatosum)**
5725. **Wattle**
5726. **Watson syndrome**
5727. **Waxy keratosis= kerinokeratosis papulosa =waxy keratosis of childhood**

5728. **Weathering nodule**
5729. **Weaver's syndrome**
5730. **Wiedemann–rautenstrauch syndrome= neonatal progeroid syndrome**
5731. **Wegener's granulomatosis**
5732. **Weightlifter's nodule**
5733. **Weill–Marchesani syndrome**
5734. **Wells syndrome =Eosinophilic cellulitis (plaque & annular & papulonodular & papulovesicular & bullous & linear & urticarial like & FDE like)**
5735. **Wende–bauckus/pegum syndrome**
5736. **Werewolf syndrome**
5737. **Werlhof's disease**
5738. **Wermer's syndrome**
5739. **Werner's syndrome**
5740. **West nile virus infection**
5741. **Weyer acrofacial dysostosis (acrodental dysostosis of weyer, curry–hall syndrome)**
5742. **Whipple disease (subcutaneous)**
5743. **Whitlow**
5744. **Whim syndrome**
5745. **Whisker Hairs**
5746. **Whitmore's disease**
5747. **White fibrous papulosis of neck**
5748. **White forelock with osteopathia striata**
5749. **White forelock with multiple malformations**
5750. **White sponge nevus of Cannon = familial white folded gingivostomatitis**
5751. **Wickham's striae**
5752. **Wiedemann-rautenstrauch syndrome**
5753. **WILD syndrome (warts, immunodeficiency, lymphedema, anogenital dysplasia)**
5754. **Williams elfin facies syndrome**
5755. **Williams–Beuren syndrome**
5756. **Winchester's syndrome**
5757. **Windmill-vane-hand syndrome**

5758. **Witkop–brearley–gentry syndrome= Hypoplastic enamel–onycholysis–hypohidrosis**
5759. **Witkop–von sallmann syndrome**
5760. **Wilson disease**
5761. **Wiskott–Aldrich syndrome**
5762. **Wnt 10 disorders**
5763. **Wolf-man syndrome**
5764. **Woolf syndrome**
5765. **Hereditary woolly hair HWH**
5766. **Woolly hair nevus**
5767. **Woolly hair syndrome**
5768. **Woolly hair-skin fragility syndrome (WHSF)**
5769. **Woolly hair, hypotrichosis, everted lower lip and outstanding ears (salamon syndrome)**
5770. **Autosomal recessive Woolly hair/hypotrichosis**
5771. **Wolf–Hirschhorn syndrome**
5772. **Wolf 's postherpetic isotopic response**
5773. **Wohlfahrtiimonas chitiniclastica**
5774. **Wong type dermatomyositis**
5775. **Woronoff's ring**
5776. **Woringer kolopp type pagetoid reticularis**
5777. **Wound**
5778. **Wrinkles**
5779. **Wrinkling skin syndrome**
5780. **Wrinkly skin syndrome**
5781. **Wyburn–mason syndrome**
5782. **Xanthelasma palpebrarum**
5783. **Xanthoma (tuberous & eruptive & tendenous & tuberoeruptive & plane)**
5784. **Cerebrotendinous Xanthomatosis**
5785. **Xanthotrichia**
5786. **Xanthomatous giant cell sarcoma**
5787. **Xeroderma pigmentosum (8 genotypes)(A,B,C,D,E,F,G,V)**
5788. **Pigmented xerodermoid**

5789. **Xeroderma pigmentosum/Cockayne syndrome complex**
5790. **Xeroderma, talipes, and enamel defect**
5791. **Xerosis cutis**
5792. **X-linked agammaglobulinemia**
5793. **X-linked hypodontia**
5794. **X-linked lymphoproliferative syndrome (duncan disease)**
5795. **X-linked reticulate pigmentary disorder=partington syndrome= Familial cutaneous amyloidosis**
5796. **X-linked severe combined immunodeficiency (SCID)**
5797. **Xmen (x-linked immunodeficiency with magnesium defect, ebv infection, and neoplasia)**
5798. **X-shaped bluish hyperpigmentation in hypohidrotic ectodermal dysplasia**
5799. **Yao syndrome**
5800. **Yaws=pian, frambesia, bouba**
5801. **Yellow fever**
5802. **Yemenite deaf-blind hypopigmentation syndrom=Warburg-Thomsen syndrome**
5803. **Yersenia pestis related diseases**
5804. **Yunis-varon syndrome**
5805. **Zanier–roubicek syndrome**
5806. **Zellweger syndrome**
5807. **Zirconium granuloma**
5808. **Zika fever**
5809. **Zimmerman –laband syndrome**
5810. **Zinser–engmann–cole syndrome**
5811. **Ziprkowski–margolis syndrome (albinism–deafness syndrome)**
5812. **Zirconium granuloma**
5813. **Zlotogora–ogur syndrome**
5814. **Zoon balanitis= plasma cell balanitis= balanitis circumscripta plasmacellularis**
5815. **Zoon vulvitis**
5816. **Zosteriform dermatoses**
5817. **Zosteriform metastasis**

5818. **Zosteriform Venous malformations =Unilateral segmental Cavernous Hemangiomatosis**
5819. **Zouboulis syndrome**
5820. **Zunich–kaye syndrome=Zunich neuroectodermal syndrome**
5821. **Zygomycosis =Mucormycosis=Phycomycosis**

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Dermatology diseases index

2025 · Hany salah soliman

طلب البحث متطابق مع محتوى داخل الكتاب

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... nevus (CMN) 3676. Bathing trunk nevus =Giant pigmented nevus 3677. Congenital panfollicular nevus 3678. Connective tissue...

CLASSIFICATION OF B-CELL LYMPHOMAS WITH PRIMARY CUTANEOUS MANIFESTATIONS	
WHO-EORTC 2005	**WHO 2008/2016**
• Primary cutaneous follicle center lymphoma	• Primary cutaneous follicle center lymphoma
• Primary cutaneous marginal zone B-cell lymphoma*	• Extranodal marginal zone lymphoma of *mucosa-associated lymphoid tissue* – MALT lymphoma
• Primary cutaneous diffuse large B-cell lymphoma, leg type	• Primary cutaneous diffuse large B-cell lymphoma, leg type
• Primary cutaneous diffuse large B-cell lymphoma, other	• Diffuse large B-cell lymphoma, NOS
• Intravascular diffuse large B-cell lymphoma	• Intravascular diffuse large B-cell lymphoma
*Includes cases previously designated as primary cutaneous immunocytoma and primary cutaneous plasmacytoma.	

Table 119.1 Classification of B-cell lymphomas with primary cutaneous manifestations – WHO-EORTC 2005 and WHO 2008/2016. NOS, not otherwise specified.

Table 1 World Health Organization/European Organization for Research and Treatment of Cancer (WHO/EORTC) classification of cutaneous lymphoma and the WHO classification (2018)

WHO-EORTC classification (2018)	WHO classification (2018)
Cutaneous T-cell and NK-cell lymphomas	**Mature T-cell and NK-cell lymphomas**
Mycosis fungoides	Mycosis fungoides
Mycosis fungoides variants and subtypes Folliculotropic Pagetoid reticulosis Granulomatous slack skin	Mycosis fungoides variants and subtypes Folliculotropic Pagetoid reticulosis Granulomatous slack skin
Sézary syndrome	Sézary syndrome
Adult T-cell leukemia/lymphoma (HTLV1$^+$)	Adult T-cell leukemia/lymphoma (HTLV1$^+$)
Primary cutaneous CD30$^+$ lymphoproliferative disorders: Lymphomatoid papulosis Primary cutaneous anaplastic large-cell lymphoma	Primary cutaneous CD30$^+$ lymphoproliferative disorders: Lymphomatoid papulosis Primary cutaneous anaplastic large-cell lymphoma
Subcutaneous panniculitis-like T-cell lymphoma	Subcutaneous panniculitis-like T-cell lymphoma
Extranodal NK/T cell lymphoma, nasal type	Extranodal NK-/T-cell lymphoma, nasal type
Primary cutaneous peripheral T-cell lymphoma Primary cutaneous aggressive epidermotropic CD8$^+$ cytotoxic T-cell lymphoma (provisional) Primary cutaneous CD4$^+$ small to medium-sized T-cell lymphoproliferative disorder (provisional) Primary cutaneous acral CD8$^+$ T-cell lymphoma (provisional) Cutaneous γ/δ T-cell lymphoma	Primary cutaneous peripheral T-cell lymphoma Primary cutaneous aggressive epidermotropic CD8$^+$ cytotoxic T-cell lymphoma (provisional) Primary cutaneous CD4$^+$ small to medium-sized T-cell lymphoproliferative disorder (provisional) Primary cutaneous acral CD8$^+$ T-cell lymphoma (provisional) Cutaneous γ/δ T-cell lymphoma
Cutaneous B-cell lymphomas	**Mature B-cell neoplasias**
Primary cutaneous follicle center lymphoma	Primary cutaneous follicle center lymphoma
Primary cutaneous marginal zone B-cell lymphoma	Primary cutaneous marginal zone B-cell lymphoma (mucosa-associated lymphoid tissue)
Primary cutaneous diffuse large B-cell lymphoma, leg type	Primary cutaneous diffuse large B-cell lymphoma, leg type Primary cutaneous diffuse large B-cell lymphoma, not otherwise specified
Intravascular large cell lymphoma	Intravascular large cell lymphoma
Epstein–Barr virus-positive mucocutaneous ulcer	Epstein–Barr virus-positive mucocutaneous ulcer
Precursor neoplasms	**Precursor neoplasms**
Blastic plasmacytoid dendritic cell neoplasm	Blastic plasmacytoid dendritic cell neoplasm

lymphomas that make the diagnosis of cutaneous lymphomas challenging. Therefore, the diagnosis is always based on a synthesis of clinical, histo- and cytomorphological, immunophenotypic, and molecular characteristics, which allow adequate classification (Kempf and Mitteldorf 2015; Stadler et al. 2013). The staging to assess the clinical spread of the disease is largely based on the tumor, node, metastasis (TNM) system used in oncology, which in its original form was, however, only conditionally suitable for cutaneous lymphomas and was therefore adapted for cutaneous lymphomas (Tables 2, 3, 4, and 5) (Kim et al. 2007; Olsen et al. 2007). Diagnostic workup is ideally performed at specialized centers and is usually interdisciplinary with oncologists, hematologists, radiologists, and dermatopathologists/hematopathologists.

The following examination is recommended if a cutaneous lymphoma is suspected:

History and Clinical Findings Skin symptoms, lymph node involvement, enlargement of tonsils, liver, and spleen.

Skin Biopsies of Adequate Size These should be partly fixed in formalin and partly frozen in liquid nitrogen. In the case of pathologically enlarged lymph nodes, lymph node biopsy should also be performed; especially in the case of high-grade malignant T- and B-cell lymphomas (exception: lymphomatoid papulosis, marginal zone lymphoma), and NK-cell lymphomas, bone marrow biopsy should be carried out.

Laboratory Tests Blood status, liver and kidney function, and lactate dehydrogenase are important. In the case of suspected leukemic variants, fluorescence-activated cell sorting (FACS) analysis (T- and B-cell subset), serum electrophoresis for B-cell lymphomas, and immune electrophoresis are recommended.

Radiological Examinations X-ray imaging of the thorax, ultrasound, and computed tomography of the liver, spleen, and lymph node stations not accessible by palpation should be carried out. Nowadays, PET-CT is often performed.

INTRODUCTION

The diagnosis and classification of cutaneous lymphomas remain one of the most challenging areas of dermatopathology. At the time the last edition of this book was published, the WHO classification of tumors of hematopoietic and lymphoid tissues based on the REAL classification of lymphomas was being published.[1,2] This classification included some cutaneous lymphomas as subgroups within the main classification framework. At the same time, the Project Group of the European Organization for Research and Treatment of Cancer published a classification of cutaneous lymphomas which came to be known as the EORTC classification of cutaneous lymphomas.[3] Those two classifications were based on a combination of histological, clinical, immunohistological, and cytogenetic features. There were certain differences in terminology and emphasis between these two classifications and the appropriateness of one or other of the classifications was debated in the literature.[4–12] The EORTC classification included only those lymphomas which appeared to be unique to the skin. The REAL/WHO classification had the advantage of including other lymphomas which involved the skin secondarily or rarely as 'primary' cutaneous lymphomas.[13] Other proposals were made for a stand-alone classification of cutaneous lymphomas based on the REAL/WHO classification.[13,14]

Recently, a consensus classification of cutaneous lymphomas, the WHO/EORTC classification of cutaneous lymphomas, has been published[15,16] and critiqued in the literature.[16–18] This compromise classification defines 'primary cutaneous lymphomas' as lymphomas that present in skin with no evidence of extracutaneous disease at the time of diagnosis. There are some inherent problems with this strict definition of primary cutaneous lymphoma as some putative primary cutaneous lymphomas are associated with systemic spread to other systems at the time of presentation, e.g. Sézary syndrome and adult T-cell leukemia/lymphoma. The definition does not encompass secondary cutaneous lymphomas. Nevertheless this classification is widely used and will be used as the framework for the discussion in this chapter (see **Table 41.1**). Other lymphoma entities which only rarely present in the skin or do so as part of systemic spread of lymphoma from another site are classified according to the general WHO classification of lymphomas.

The current diagnosis of lymphomas requires some knowledge of lymphocyte ontogeny and techniques which are used to demonstrate such aspects as cell phenotype, clonality of a proliferation, and cytogenetic features including the presence of viral genetic material. Most laboratories are now able to perform these techniques, including immunohistochemistry, in-situ hybridization, polymerase chain reaction technology (PCR), and fluorescence in-situ hybridization (FISH).[19–22] Flow cytometry is also used for immunophenotyping[23] but

Table 41.1 The Classification of cutaneous lymphomas used in this chapter (based on the WHO–EORTC classification)

Cutaneous T-cell and NK-cell lymphomas
Mycosis fungoides and subtypes
Folliculotropic mycosis fungoides
Pagetoid reticulosis
Granulomatous slack skin
Hydroa vacciniforme-like lymphoma
Sézary syndrome
Adult T-cell leukemia/lymphoma
Primary cutaneous $CD30^+$ lymphoproliferative disorders
Primary cutaneous anaplastic large cell lymphoma
Lymphomatoid papulosis
Subcutaneous panniculitis-like T-cell lymphoma
Extranodal NK/T-cell lymphoma, nasal type
Primary cutaneous peripheral T-cell lymphoma, unspecified
Primary cutaneous aggressive epidermotropic $CD8^+$ T-cell lymphoma (provisional)
Cutaneous γ/δ T-cell lymphoma (provisional)
Primary cutaneous $CD4^+$ small/medium pleomorphic T-cell lymphoma (provisional)
Cutaneous B-cell lymphomas
Primary cutaneous marginal zone B-cell lymphoma
Primary cutaneous follicle center cell lymphoma
Primary cutaneous diffuse large B-cell lymphoma, leg type
Primary cutaneous diffuse large B-cell lymphoma, other
Intravascular large B-cell lymphoma
Plasmablastic lymphoma
T-cell/histiocyte-rich B-cell lymphoma
Lymphomatoid granulomatosis
Precursor hematologic neoplasm
Blastic plasmacytoid dendritic cell neoplasm
Other T/NK-cell lymphomas that may involve the skin
Precursor T-lymphoblastic lymphoma/leukemia
T-cell prolymphocytic leukemia
Angioimmunoblastic T-cell lymphoma
Primary systemic anaplastic large cell lymphoma
Intravascular T- and NK-cell lymphoma
Aggressive NK-cell leukemia
Other T/NK-cell lymphomas and leukemias
Other B-cell lymphomas that may involve the skin
Precursor B-lymphoblastic leukemia/lymphoma
Chronic lymphocytic leukemia/small lymphocytic lymphoma
Mantle cell lymphoma
Primary effusion lymphoma
Lymphoplasmacytic lymphoma/Waldenström's macroglobulinemia
Burkitt and Burkitt-like lymphoma
Plasmacytoma and secondary myeloma
Other lymphomas
Hodgkin lymphoma
Cutaneous infiltrates of leukemias
Myeloid leukemias, myeloproliferative diseases and myelodysplastic syndromes
Lymphoid hyperplasia mimicking primary lymphoma
Lymphoid hyperplasia simulating B-cell lymphoma
Lymphomatoid drug reactions
Reactions resembling $CD30^+$ lymphoproliferative disorders
Pseudolymphomatous folliculitis
Jessner's lymphocytic infiltrate
Acral pseudolymphomatous angiokeratoma
Cutaneous $CD8^+$ T-cell infiltrates in HIV/AIDS
Cutaneous infiltrates in post-transplant lymphoproliferative disorders
Other lymphoproliferative disorders associated with immunosuppression
Methotrexate-associated lymphoproliferative disorders
Cutaneous infiltrates in HIV/AIDS
Miscellaneous
Extramedullary hematopoiesis

Table 1.2 Revised 2018 WHO–EORTC classification of primary cutaneous lymphomas

Cutaneous T-cell lymphomas
Mycosis fungoides
Folliculotropic mycosis fungoides
Pagetoid reticulosis
Granulomatous slack skin
Sézary syndrome
Adult T-cell leukemia/lymphoma
Primary cutaneous CD30$^+$ lymphoproliferative disorders
Cutaneous anaplastic large cell lymphoma
Lymphomatoid papulosis
Subcutaneous panniculitis-like T-cell lymphoma
Extranodal NK/T-cell lymphoma, nasal type
Chronic active EBV infection
Primary cutaneous peripheral T-cell lymphoma, rare subtypes
Primary cutaneous γ/δ T-cell lymphoma
CD8$^+$ aggressive epidermotropic cytotoxic T-cell lymphoma
Primary cutaneous CD4$^+$ small/medium T-cell lymphoproliferative disorder
Primary cutaneous acral CD8$^+$ T-cell lymphoma
Primary cutaneous peripheral T-cell lymphoma, not otherwise specified
Cutaneous B-cell lymphomas
Primary cutaneous marginal zone lymphoma
Primary cutaneous follicle center lymphoma
Primary cutaneous diffuse large cell lymphoma, leg type
EBV$^+$ mucocutaneous ulcer
Intravascular large B-cell lymphoma

Adapted from Willemze *et al.* [2] (American Society of Hematology). Provisional entities are in italic.

Table 1. WHO/EORTC classification of cutaneous lymphomas

WHO/EORTC classification of cutaneous lymphomas	WHO classification for lymphoid tissues† (ICD-O Code)
Cutaneous T-cell and NK-cell lymphomas	Mature T-cell and NK-cell neoplasms
Mycosis fungoides (MF)	Mycosis fungoides (MF) (9700/3)
MF variants and subtypes	MF variants and subtypes
Folliculotropic MF	Folliculotropic MF
Pagetoid reticulosis	Pagetoid reticulosis
Granulomatous slack skin	Granulomatous slack skin
Sézary syndrome	Sézary syndrome (9701/3)
Adult T-cell leukaemia/lymphoma	Adult T-cell leukaemia/lymphoma (9827/3)
Primary cutaneous CD30+ lymphoproliferative disorders	Primary cutaneous CD30+ T-cell lymphoproliferative disorders
Primary cutaneous anaplastic large cell lymphoma	Primary cutaneous anaplastic large cell lymphoma (9718/3)
Lymphomatoid papulosis	Lymphomatoid papulosis (9718/1)
Subcutaneous panniculitis-like T-cell lymphoma*	Subcutaneous panniculitis-like T-cell lymphoma* (9708/3)
Extranodal NK/T-cell lymphoma, nasal type	Extranodal NK/T-cell lymphoma, nasal type (9719/3)
Primary cutaneous peripheral T-cell lymphoma, unspecified	Primary cutaneous peripheral T-cell lymphoma, rare subtypes
Primary cutaneous aggressive epidermotropic CD8+ T-cell lymphoma (provisional)	Primary cutaneous CD8+ aggressive epidermotropic cytotoxic T-cell lymphoma (provisional) (9709/3)
Cutaneous γ/δ+ T-cell lymphoma (provisional)	Primary cutaneous γ/δ T-cell lymphoma (9726/3)‡
Primary cutaneous CD4+ small/medium-sized pleomorphic T-cell lymphoma (provisional)	Primary cutaneous CD4+ small/medium T-cell lymphoma (provisional) (9709/3)
Cutaneous B-cell lymphomas	Mature B-cell neoplasms
Primary cutaneous marginal zone B-cell lymphoma	Extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma) (9699/3)
Primary cutaneous immunocytoma	
Primary cutaneous plasmacytoma	
Follicular hyperplasia with monotypic plasma cells	
Primary cutaneous follicle centre lymphoma	Primary cutaneous follicle centre lymphoma (9597/3)
Growth patterns: follicular, follicular and diffuse, diffuse	Diffuse large B-cell lymphoma, NOS (9680/3)
Primary cutaneous diffuse large B-cell lymphoma, leg type	Primary cutaneous diffuse large B-cell lymphoma, leg type (9680/3)
Primary cutaneous diffuse large B-cell lymphoma, other	Intravascular large B-cell lymphoma (9712/3)
Primary cutaneous intravascular large B-cell lymphoma	
Precursor haematological neoplasm	Precursor neoplasms
CD4+/CD56+ haematodermic neoplasm (formerly blastic NK cell lymphoma)	Blastic plasmacytoid dendritic cell neoplasm (9727/3)

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Table 1.1 Revised 4th edition of the WHO *Classification of Tumours of Haematopoietic and Lymphoid Tissues* (2017)

Myeloproliferative neoplasms
Chronic myeloid leukemia, *BCR-ABL1*–positive
Chronic neutrophilic leukemia
Polycythemia vera
Primary myelofibrosis
Essential thrombocythemia
Chronic eosinophilic leukemia, NOS
Myeloproliferative neoplasm, unclassifiable

Mastocytosis
Cutaneous mastocytosis
Systemic mastocytosis
Indolent systemic mastocytosis
Mast cell sarcoma

Myeloid/lymphoid neoplasms with eosinophilia and gene rearrangement
Myeloid/lymphoid neoplasms with *PDGFRA* rearrangement
Myeloid/lymphoid neoplasms with *PDGFRB* rearrangement
Myeloid/lymphoid neoplasms with *FGFR1* rearrangement
Myeloid/lymphoid neoplasms with PCM1-JAK2 9968/3

Myelodysplastic/myeloproliferative neoplasms
Chronic myelomonocytic leukemia
Atypical chronic myeloid leukemia, *BCR-ABL1*–negative
Juvenile myelomonocytic leukemia
Myelodysplastic/myeloproliferative neoplasm with ring sideroblasts and thrombocytosis
Myelodysplastic/myeloproliferative neoplasm, unclassifiable

Myelodysplastic syndromes
Myelodysplastic syndrome with single lineage dysplasia
Myelodysplastic syndrome with ring sideroblasts and single lineage dysplasia
Myelodysplastic syndrome with ring sideroblasts and multilineage dysplasia
Myelodysplastic syndrome with multilineage dysplasia
Myelodysplastic syndrome with excess blasts
Myelodysplastic syndrome with isolated del(5q)
Myelodysplastic syndrome, unclassifiable
Refractory cytopenia of childhood (provisional entity)

Myeloid neoplasms with germline predisposition
Acute myeloid leukemia with germline *CEBPA* mutation
Myeloid neoplasms with germline *DDX41* mutation
Myeloid neoplasms with germline *RUNX1* mutation
Myeloid neoplasms with germline *ANKRD26* mutation
Myeloid neoplasms with germline *ETV6* mutation
Myeloid neoplasms with germline *GATA2* mutation

Acute myeloid leukemia (AML) and related precursor neoplasms

AML with recurrent genetic abnormalities
AML with t(8;21)(q22;q22.1); *RUNX1-RUNX1T1*
AML with inv.(16)(p13.1q22) or t(16;16)(p13.1;q22); *CBFB-MYH11*
Acute promyelocytic leukemia with *PML-RARA*
AML with t(9;11)(p21.3;q23.3); *KMT2A-MLLT3*
AML with t(6;9)(p23;q34.1); *DEK-NUP214*
AML with inv.(3)(q21.3q26.2) or t(3;3)(q21.3;q26.2); *GATA2*, *MECOM*
AML (megakaryoblastic) with t(1;22)(p13.3;q13.1); *RBM15-MKL1*
AML with BCR-ABL1 (provisional entity)
AML with mutated NPM1
AML with biallelic mutation of *CEBPA*
AML with mutated RUNX1 (provisional entity)

AML with myelodysplasia-related changes

Therapy-related myeloid neoplasms

Acute myeloid leukaemia, NOS
AML with minimal differentiation
AML without maturation
AML with maturation
Acute myelomonocytic leukemia
Acute monoblastic and monocytic leukemia
Pure erythroid leukemia
Acute megakaryoblastic leukemia
Acute basophilic leukemia
Acute panmyelosis with myelofibrosis

Myeloid sarcoma (may present primary in the skin)

Myeloid proliferations associated with Down syndrome
Transient abnormal myelopoiesis associated with Down syndrome
Myeloid leukemia associated with Down syndrome

Blastic plasmacytoid dendritic cell neoplasm (often presents primary in the skin)

Acute leukemias of ambiguous lineage
Acute undifferentiated leukemia
Mixed-phenotype acute leukemia with t(9;22)(q34.1;q11.2); *BCR-ABL1*
Mixed-phenotype acute leukemia with t(v;11q23.3); *KMT2A*-rearranged
Mixed-phenotype acute leukemia, B/myeloid, NOS
Mixed-phenotype acute leukemia, T/ myeloid, NOS
Mixed-phenotype acute leukemia, NOS, rare types
Acute leukemias of ambiguous lineage, NOS

Precursor lymphoid neoplasms
B-lymphoblastic leukemia/lymphoma, NOS (may present primary in the skin)
B-lymphoblastic leukemia/lymphoma with recurrent genetic abnormalities
B-lymphoblastic leukemia/lymphoma with t(9;22)(q34.1;q11.2); *BCR-ABL1*
B-lymphoblastic leukemia/lymphoma with t(v;11q23.3); *KMT2A*-rearranged
B-lymphoblastic leukemia/lymphoma with t(12;21)(p13.2;q22.1); *ETV6-RUNX1*
B-lymphoblastic leukemia/lymphoma with hyperdiploidy
B-lymphoblastic leukemia/lymphoma with hypodiploidy (hypodiploid ALL)
B-lymphoblastic leukemia/lymphoma with t(5;14)(q31.1;q32.1); IGH/*IL3*
B-lymphoblastic leukemia/lymphoma with t(1;19)(q23;p13.3); *TCF3-PBX1*
B-lymphoblastic leukemia/lymphoma, BCR-ABL1–like (provisional entity)
B-lymphoblastic leukemia/lymphoma with iAMP21
T-lymphoblastic leukemia/lymphoma
Early T-cell precursor acute lymphoblastic leukemia
NK-lymphoblastic leukemia/lymphoma (provisional entity)

Mature B-cell neoplasms
Chronic lymphocytic leukemia (CLL)/small lymphocytic lymphoma
Monoclonal B-cell lymphocytosis
  CLL type
  Non-CLL type
B-cell prolymphocytic leukemia
Splenic marginal zone lymphoma
Hairy cell leukemia
Splenic B-cell lymphoma/leukemia, unclassifiable
  Splenic diffuse red pulp small B-cell lymphoma (provisional entity)
  Hairy cell leukemia variant (provisional entity)
Lymphoplasmacytic lymphoma
Monoclonal gammopathy of undetermined significance, IgM
Heavy chain diseases
  μ Heavy chain disease
  γ Heavy chain disease
  α Heavy chain disease
Plasma cell neoplasms
  Non-IgM monoclonal gammopathy of undetermined significance
  Plasma cell myeloma
    Plasma cell myeloma variants
Plasmacytoma
  Solitary plasmacytoma of bone
  Extraosseous plasmacytoma
Monoclonal immunoglobulin deposition diseases
  Primary amyloidosis

Table 1.1 (*cont'd*)

- Light chain and heavy chain deposition diseases
- Plasma cell neoplasms with associated paraneoplastic syndrome
  - POEMS syndrome
  - TEMPI syndrome (provisional entity)
- *Extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma) (includes primary cutaneous marginal zone lymphoma)*
- Nodal marginal zone lymphoma
  - Pediatric nodal marginal zone lymphoma
- Follicular lymphoma
  - In situ follicular neoplasia
  - Duodenal-type follicular lymphoma 9690/3
- Pediatric-type follicular lymphoma
- Large B-cell lymphoma with IRF4 rearrangement (provisional entity)
- *Primary cutaneous follicle center lymphoma*
- Mantle cell lymphoma
  - Leukemic non-nodal mantle cell lymphoma
  - In situ mantle cell neoplasia
- Diffuse large B-cell lymphoma (DLBCL), NOS
  - Germinal center B-cell subtype
  - Activated B-cell subtype
- T-cell/histiocyte-rich large B-cell lymphoma
- Primary DLBCL of the CNS
- *Primary cutaneous DLBCL, leg type*
- EBV-positive DLBCL, NOS
- *EBV-positive mucocutaneous ulcer*
- DLBCL associated with chronic inflammation
- Lymphomatoid granulomatosis
  - Grade 1 or 2
  - Grade 3
- Primary mediastinal (thymic) large B-cell lymphoma
- *Intravascular large B-cell lymphoma (often presents primary in the skin)*
- ALK-positive large B-cell lymphoma
- Plasmablastic lymphoma
- Primary effusion lymphoma
- HHV8-associated lymphoproliferative disorders
  - Multicentric Castleman disease
  - HHV8-positive DLBCL, NOS
  - HHV8-positive germinotropic lymphoproliferative disorder
- Burkitt lymphoma
- Burkitt-like lymphoma with 11q aberration (provisional entity)
- High-grade B-cell lymphoma
- High-grade B-cell lymphoma with *MYC* and *BCL2* and/or *BCL6* rearrangement
- High-grade B-cell lymphoma, NOS
- B-cell lymphoma, unclassifiable, with features intermediate between DLBCL and classic Hodgkin lymphoma

Mature T- and NK-cell neoplasms

- T-cell prolymphocytic leukemia
- T-cell large granular lymphocytic leukemia
- Chronic lymphoproliferative disorder of NK cells (provisional entity)
- Aggressive NK-cell leukemia
- EBV-positive T-cell lymphoproliferative diseases of childhood
  - Systemic EBV-positive T-cell lymphoma of childhood
  - Chronic active EBV infection of T- and NK-cell type, systemic form
  - *Hydroa vacciniforme-like lymphoproliferative disorder*
  - *Severe mosquito bite allergy*
- *Adult T-cell leukemia/lymphoma (may present primary in the skin)*
- *Extranodal NK/T-cell lymphoma, nasal type (may present primary in the skin)*
- Intestinal T-cell lymphoma
  - Enteropathy-associated T-cell lymphoma
  - Monomorphic epitheliotropic intestinal T-cell lymphoma
  - Intestinal T-cell lymphoma, NOS
  - Indolent T-cell lymphoproliferative disorder of the gastrointestinal tract (provisional entity)
- Hepatosplenic T-cell lymphoma
- *Subcutaneous panniculitis-like T-cell lymphoma*
- *Mycosis fungoides*
- *Sézary syndrome*
- *Primary cutaneous CD30-positive T-cell lymphoproliferative disorders*
  - *Lymphomatoid papulosis*
  - *Primary cutaneous anaplastic large cell lymphoma*
- *Primary cutaneous peripheral T-cell lymphomas, rare subtypes*
  - *Primary cutaneous gamma delta T-cell lymphoma*
  - *Primary cutaneous CD8-positive aggressive epidermotropic cytotoxic T-cell lymphoma (provisional entity)*
  - *Primary cutaneous acral CD8-positive T-cell lymphoma (provisional entity)*
  - *Primary cutaneous CD4-positive small/medium T-cell lymphoproliferative disorder (provisional entity)*
- *Peripheral T-cell lymphoma, NOS (may present primary in the skin)*
- **Angioimmunoblastic T-cell lymphoma and other nodal lymphomas of T follicular helper cell origin**
  - Angioimmunoblastic T-cell lymphoma
  - Follicular T-cell lymphoma
  - Nodal peripheral T-cell lymphoma with T follicular helper phenotype
- Anaplastic large cell lymphoma, ALK-positive
- Anaplastic large cell lymphoma, ALK-negative
- Breast implant-associated anaplastic large cell lymphoma (provisional entity)

Hodgkin lymphomas

- Nodular lymphocyte-predominant Hodgkin lymphoma
- Classic Hodgkin lymphoma
  - Nodular sclerosis classic Hodgkin lymphoma
  - Lymphocyte-rich classic Hodgkin lymphoma
  - Mixed cellularity classic Hodgkin lymphoma
  - Lymphocyte-depleted classic Hodgkin lymphoma

Immunodeficiency-associated lymphoproliferative disorders (may present primary in the skin)

- Lymphoproliferative diseases associated with primary immune disorders
- Lymphomas associated with HIV infection
- Posttransplant lymphoproliferative disorders (PTLDs)
  - Nondestructive PTLDs
  - Plasmacytic hyperplasia PTLD
  - Infectious mononucleosis PTLD
  - Florid follicular hyperplasia PTLD
  - Polymorphic PTLD
  - Monomorphic PTLDs (B- and T/NK-cell types)
  - Classic Hodgkin lymphoma PTLD
  - Other iatrogenic immunodeficiency-associated lymphoproliferative disorders

Histiocytic and dendritic cell neoplasms (may present primary in the skin)

- Histiocytic sarcoma
- Tumors derived from Langerhans cells
  - Langerhans cell histiocytosis
  - Langerhans cell sarcoma
- Indeterminate dendritic cell tumor
- Interdigitating dendritic cell sarcoma
- Follicular dendritic cell sarcoma
  - Inflammatory pseudotumor-like follicular/fibroblastic dendritic cell sarcoma
- Fibroblastic reticular cell tumor
- Disseminated juvenile xanthogranuloma
- Erdheim–Chester disease

Italic, primary cutaneous lymphomas.
Adapted from Swerdlow *et al*. [1].

Table 1.2 The WHO classification of tumours of haematopoietic and lymphoid tissues.

B-cell neoplasms
Precursor B-cell neoplasms
Precursor B-lymphoblastic leukaemia–lymphoma

Mature B-cell neoplasms
Chronic lymphocytic leukaemia/small lymphocytic lymphoma
B-cell prolymphocytic leukaemia
Lymphoplasmacytic lymphoma
Splenic marginal zone lymphoma
Hairy cell leukaemia
Plasma cell myeloma
Solitary plasmacytoma of bone
Extraosseous plasmacytoma
Extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma)
Nodal marginal zone B-cell lymphoma
Follicular lymphoma
Mantle cell lymphoma
Diffuse large B-cell lymphoma
Mediastinal (thymic) large B-cell lymphoma
Intravascular large B-cell lymphoma
Primary effusion lymphoma
Burkitt lymphoma/leukaemia

B-cell proliferations of uncertain malignant potential
Lymphomatoid granulomatosis
Post-transplant lymphoproliferative disorder, polymorphic

T- and NK-cell neoplasms
Precursor T-cell neoplasms
Precursor T-lymphoblastic leukaemia/lymphoma
Blastic NK-cell lymphoma (neoplasm of uncertain lineage and stage of differentiation)

Mature T- and NK-cell neoplasms
T-cell prolymphocytic leukaemia
T-cell large granular lymphocytic leukaemia
Aggressive NK-cell leukaemia
Adult T-cell leukaemia/lymphoma
Extranodal NK/T-cell lymphoma, nasal type
Enteropathy-type T-cell lymphoma
Hepatosplenic T-cell lymphoma
Subcutaneous panniculitis-like T-cell lymphoma
Mycosis fungoides
Sézary syndrome
Anaplastic large cell lymphoma, primary cutaneous
Peripheral T-cell lymphoma, unspecified
Angioimmunoblastic T-cell lymphoma
Anaplastic large cell lymphoma

T-cell proliferation of uncertain malignant potential
Lymphomatoid papulosis

Hodgkin lymphoma
Nodular lymphocyte-predominant Hodgkin lymphoma
Nodular sclerosis classical Hodgkin lymphoma
Lymphocyte-rich classical Hodgkin lymphoma
Mixed cellularity classical Hodgkin lymphoma
Lymphocyte-depleted classical Hodgkin lymphoma

may rarely present a monoclonal pattern, and a 'germline' or polyclonal pattern may be observed in clear-cut lymphomas. In addition, lack of sensitivity may give rise to negative results in many cases of early cutaneous T- or B-cell lymphoma.

A common pitfall in molecular analysis of cutaneous lymphoproliferative disorders is the presence of 'pseudomonoclonal' rearrangement of the J_H gene in cutaneous lymphoid infiltrates characterized by the presence of only a few B lymphocytes. In fact, it is not uncommon to observe this phenomenon in lesions of cutaneous T-cell lymphoma with reactive B lymphocytes. A good rule is to perform at least two (better three) separate extractions of DNA from different sections of the biopsy tissue, and to run one independent PCR assay for every extraction. In this way, different 'pseudomonoclonal' bands will be observed, thus showing overall a polyclonal pattern.

Molecular analyses are also widely used in haematology to identify particular genetic aberrations characteristic of a specific clinicopathological entity, such as the interchromosomal 14;18 translocation typical of nodal follicular lymphoma or the t(2;5) of nodal anaplastic large cell lymphoma. Unfortunately, as yet only a few specific genetic alterations have been identified in primary cutaneous lymphomas.

Other methods in the study of cutaneous lymphoid infiltrates

Fluorescence *in situ* hybridization technique

Chromosomal abnormalities are becoming increasingly important in the study and classification of haematopoietic neoplasms. In recent years, new methods have been developed for evaluation of tissue specimens fixed in formalin and embedded in paraffin, allowing the investigation of routine biopsy samples and archive material. The fluorescence *in situ* hybridization (FISH) technique is based on the same principle as the Southern blot method, relying on the annealing of single-stranded DNA to complementary DNA.

Depending on the probes selected, the FISH method can be used to detect different types of chromosomal abnormalities, including monosomy, trisomy and other aneuploidies, as well as translocations and deletions. The use of FISH applied to routinely processed tissue specimens identified a hitherto unknown t(14;18)(q32;q21) involving the J_H and MALT1 genes in a subset of cases of primary cutaneous marginal zone B-cell lymphomas [15].

Microarrays

The recent development of techniques of comparative genomic hybridization, and the identification of over 19 000 human genes by the Human Genome Project, allows one to

Table 1 WHO classification of mature B-cell, T-cell and NK-cell neoplasms (2008)

Precursor lymphoid neoplasms
B lymphoblastic leukaemia/lymphoma NOS
B lymphoblastic leukaemia/lymphoma with recurrent genetic abnormalities
B lymphoblastic leukaemia/lymphoma with t(9;22); bcr-abl1
B lymphoblastic leukaemia/lymphoma with t(v;11q23); MLL rearranged
B lymphoblastic leukaemia/lymphoma with t(12:21); TEL-AML1 & ETV6-RUNX1
B lymphoblastic leukaemia/lymphoma with hyperploidy
B lymphoblastic leukaemia/lymphoma with hypodiploidy
B lymphoblastic leukaemia/lymphoma with t(5;14); IL3-IGH
B lymphoblastic leukaemia/lymphoma with t(1;19); E2A-PBX1 & TCF3-PBX1
T lymphoblastic leukaemia/lymphoma
Mature B-cell neoplasms
Chronic lymphocytic leukaemia/small lymphocytic lymphoma
B-cell prolymphocytic leukaemia
Splenic marginal zone lymphoma
Hairy cell leukaemia
Lymphoplasmacytic lymphoma/Waldenström macroglobulinaemia
Heavy chain disease
Plasma cell myeloma
Solitary plasmacytoma of bone
Extraosseous plasmacytoma
Extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue (MALT) type
Nodal marginal zone lymphoma
Follicular lymphoma
Primary cutaneous follicular lymphoma
Mantle cell lymphoma
Diffuse large B-cell lymphoma, NOS
(T-cell/histiocyte-rich type; primary CNS type; primary leg skin type and EBV + elderly type)
Diffuse large B-cell lymphoma with chronic inflammation
Lymphomatoid granulomatosis
Primary mediastinal large B-cell lymphoma
Intravascular large B-cell lymphoma
ALK + large B-cell lymphoma
Plasmablastic lymphoma
Large B-cell lymphoma associated with HHV8 + Castleman disease
Primary effusion lymphoma
Burkitt's lymphoma
B-cell lymphoma, unclassifiable, Burkitt's-like
B-cell lymphoma, unclassifiable, Hodgkin's lymphoma-like
Mature T- and NK-cell neoplasms
T-cell prolymphocytic leukaemia
T-cell large granular lymphocytic leukaemia
Chronic lymphoproliferative disorder of NK cells
Aggressive NK cell leukaemia
Systemic EBV + T-cell lymphoproliferative disorder of childhood
Hydroa vacciniforme-like lymphoma
Adult T-cell lymphoma/leukaemia
Extranodal NK/T-cell lymphoma, nasal type
Enteropathy-associated T-cell lymphoma
Hepatosplenic T-cell lymphoma
Subcutaneous panniculitis-like T-cell lymphoma
Mycosis fungoides
Sézary syndrome
Primary cutaneous CD30 + T-cell lymphoproliferative disorder
Primary cutaneous gamma/delta-positive T-cell lymphoma
Peripheral T-cell lymphoma, NOS
Angioimmunoblastic T-cell lymphoma
Anaplastic large cell lymphoma, ALK + type
Anaplastic large cell lymphoma, ALK-type
Hodgkin's lymphoma (Hodgkin's disease)
Nodular lymphocyte-predominant Hodgkin's lymphomas
Classical Hodgkin's lymphoma
Nodular sclerosis Hodgkin's lymphoma
Lymphocyte-rich classical Hodgkin's lymphoma
Mixed cellularity Hodgkin's lymphoma
Lymphocyte depletion Hodgkin's lymphoma
Posttransplant lymphoproliferative disorders (PTLD)
Plasmacytic hyperplasia
Infectious mononucleosis-like PTLD
Polymorphic PTLD
Monomorphic PTLD (B and NK/T-cell types)
Classical HD-type PTLD
Histiocytic and dendritic cell neoplasms
Histiocytic sarcoma
Langerhans cell histiocytosis
Langerhans cell sarcoma
Interdigitating dendritic cell sarcoma
Follicular dendritic cell sarcoma
Fibroblastic reticular cell tumour
Indeterminate dendritic cell sarcoma
Disseminated juvenile xanthogranuloma

small clonal lymphoid populations and the identification of diseases characterised by the involvement of specific anatomical sites or by other clinical features, such as age (Jaffe *et al*, 2008).

Non-Hodgkin's lymphoma (NHL) is the most frequent type of tumour of the head and neck, representing about 75% of lymphomas in this area (Boring *et al*, 1993). Some types of NHL are especially common in this area. The common B-cell neoplasms are precursor B-lymphoblastic leukaemia/lymphoma, chronic lymphocytic leukaemia/small lymphocytic lymphoma, extranodal marginal-zone B-cell lymphoma of MALT, follicular lymphoma, mantle cell lymphoma, diffuse large B-cell lymphoma, and Burkitt's lymphoma/leukaemia. The most frequent T-cell and NK cell NHL is the extranodal NK/T-cell lymphoma, nasal type (Harris *et al*, 1994).

Clinical evaluation

The timing of diagnosis differs considerably depending on the type of lymphoma. HL is diagnosed more frequently in patients 20–30 years of age, whereas NHL is diagnosed more frequently between 70 and 80 years of age (Urquhart and Berg, 2001). In childhood, extranodal NHL may mimic other clinical entities, such as inflammatory disease, polymorphic reticulosis or rhabdomyosarcoma (La Quaglia, 1994). There are no gender differences between HL and NHL in the head and neck. Generalised symptoms, such as fever, weight loss, night sweats, fatigue or pruritus (Urquhart and Berg, 2001), are more frequent in HL than NHL (41% *vs* 27% respectively). Thus, taking into account the minor frequency of HL in the head and neck, systemic symptoms are not common in this area.

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Table 1 Classification of Leukemias

French–American–British (FAB) classification of acute leukemias
M0 Acute myeloid leukemia with minimal evidence of myeloid differentiation
M1 Acute myeloid leukemia without maturation
M2 Acute myeloid leukemia with maturation
M3 Acute promyelocytic leukemia
M4 Acute myelomonocytic leukemia
M5 Acute monocytic leukemia
M6 Acute erythroleukemia
M7 Acute megakaryoblastic leukemia
Acute lymphoblastic leukemias
Chronic leukemias
Chronic myeloid leukemia
Chronic lymphocytic leukemia, B-cell type
Chronic lymphocytic leukemia, T-cell type
T-cell prolymphocytic leukemia
Adult T-cell leukemia/lymphoma
Hairy cell leukemia
Chronic myelomonocytic leukemia

Table 23.1 Classification of precursor lymphoid neoplasms according to the WHO [17]

B-lymphoblastic leukemia/lymphoma, NOS
B-lymphoblastic leukemia/lymphoma with recurrent genetic abnormalities
B-lymphoblastic leukemia/lymphoma with t(9;22)(q34;q11.2); *BCR-ABL1*
B-lymphoblastic leukemia/lymphoma with t(v;11q23); *MLL* rearranged
B-lymphoblastic leukemia/lymphoma with t(12;21)(p13;q22); *TEL-AML1 (ETV6-RUNX1)*
B-lymphoblastic leukemia/lymphoma with hyperdiploidy
B-lymphoblastic leukemia/lymphoma with hypodiploidy (hypodiploid ALL)
B-lymphoblastic leukemia/lymphoma with t(5;14)(q31;q32); *IL3-IGH*
B-lymphoblastic leukemia/lymphoma with t(1;19)(q23;p13.3); *E2A-PBX1 (TCF3-PBX1)*
T-lymphoblastic leukemia/lymphoma

Table 2.4 Historical terms used for mycosis fungoides*

Erythrodermie pityriasique en plaques disseminées
Parakeratosis variegata
Parapsoriasis en plaques
Parapsoriasis lichenoides
Parapsoriasis variegata
Poikiloderma vasculare atrophicans
Premycotic erythema
Prereticulotic poikiloderma
Retiform type of parapsoriasis
Xanthoerythroderma perstans

* Some of these terms are still used today in some centers.

TABLE 2 Classification of cutaneous pseudolymphomas

Nodular pseudolymphomas
Nodular B-cell pseudolymphoma
Borrelia-associated nodular B-cell pseudolymphoma
Nodular T-cell and mixed pseudolymphoma
Nodular CD30+ pseudolymphoma
Pseudolymphomas as simulators of mycosis fungoides or of other CTCLs
Lymphomatoid contact dermatitis
Lymphomatoid drug reaction
Actinic reticuloid
CD8+ T-cell pseudolymphoma in immunodeficiency
Infections as simulators of T-cell lymphomas
Borrelia-associated T-cell pseudolymphoma
Leishmaniasis-associated T-cell pseudolymphoma[a]
Herpesvirus-associated T-cell pseudolymphoma
Syphilis-associated pseudolymphoma
Other Infections als simulators of CTCL
Inflammatory dermatosis as simulators of CTCL
Other pseudolymphomas
T-cell rich angiomatoid pseudolymphoma
Acral pseudolymphomatous angiokeratoma (APA)
T-cell-rich angiomatoid polypoid pseudolymphoma (TRAPP)
Primary cutneous angioplasmocellular hyperplasia
Lymphoplasmacytoid plaque (LPP)
Cutaneous plasmocytosis
Intravascular pseudolymphomas
Benign atypical intravascular (CD30+) lymphoproliferation

[a]Also presents as nodular T-cell pseudolymphoma.

Predominant T-cell pseudolymphomas
Drug induced • Localized (nodular histology, predominantly CD4) • Diffuse (band-like histology)
UV-induced: for example, chronic actinic dermatitis (predominantly CD8, or mixed)
HIV-related (band-like histology, CD8+)
Insect bite reactions
Idiopathic T-cell CPL (nodular or band-like CD4+)
Predominant B cell pseudolymphomas
Borrelia-related cutaneous lymphoid hyperplasia
Varicella-zoster virus-related CPL
Vaccination-induced CLH
CLH secondary to insect bites
Drug-induced (rare)
Idiopathic

TABLE 1 | Isolated non-syndromic hereditary hair loss disorders, classified by OMIM naming system.

OMIM Name	Disease	Inheritance	Gene and locus	Phenotype
Hypotrichosis 1 (HYPT1)	Hereditary hypotrichosis simplex type 1 (HHS1)	AD	APCDD1 18p11.22	Normal hair at birth, generalised hypotrichosis progressing from early childhood, with sparse, thin and short hair on scalp and body. Sparing of eyebrow, eyelash and beard hair [4].
Hypotrichosis 2 (HYPT2)	Hereditary hypotrichosis simplex type 2 (HHS2)	AD	CDSN 6p21.3	Normal hair at birth, progressive scalp hair loss from the first decade to diffuse loss by the third decade. Sparse, fine, short scalp hairs. Sparing of eyebrow, eyelash, beard and axillary hair [5].
Hypotrichosis 3 (HYPT3)	Dominant hereditary hypotrichosis 3	AD	KRT74 12q13	Sparse scalp hair from birth, sometimes wiry irregular hair in childhood, with progressive loss to puberty. Sparing of eyebrow, eyelash and facial hair. Normal teeth and nails [6].
Hypotrichosis 4 (HYPT4)	Marie Unna hereditary hypotrichosis 1 (MUHH1)	AD	U2HR 8p21.2	Sparse or absent hair at birth. Slow-growing, variable, coarse, wiry, fragile hair from childhood. Affecting scalp, eyebrows and body hair. Electron microscopy displaying an irregular twisting hair dystrophy and longitudinal grooving [7].
Hypotrichosis 5 (HYPT5)	Marie Unna hereditary hypotrichosis 2 (MUHH2)	AD	EPS8L3 1p13.3	Sparse to absent scalp hair at birth. Wiry, irregular hair in childhood. Thin eyebrows and eyelashes with absence of axillary and pubic hair [8].
Hypotrichosis 6 (HYPT6)	Localised autosomal recessive hypotrichosis 1 (LAH1)	AR	DSG4 18q12	Sparse fragile scalp hairs, affected or absent eyebrows and eyelashes and sparing facial, axillary and pubic hairs. Occasionally hyperkeratotic follicular papules on scalp or other affected areas—an association among LAH phenotypes [9].
Hypotrichosis 7 (HYPT7)	Localised autosomal recessive hypotrichosis 2 (LAH2)	AR	LIPH 3q27.2	Sparse, wiry and twisted to woolly scalp hair. Wide spectrum of body hair phenotype, with individuals affected to different degrees in adulthood [10].
Hypotrichosis 8 (HYPT8)	Localised autosomal recessive hypotrichosis 3 (LAH3)	AR	LPAR6 13q14	Tightly curled, woolly, fragile and slow-growing scalp and trunk hair, and normal to sparse eyebrows and eyelashes [9, 11]. Sparing of beard, and axillary and pubic hairs [11].
Hypotrichosis 9 (HYPT9)	Localised autosomal recessive hypotrichosis 4 (LAH4)	AR	Unknown 10q11.23-q22.3	Thin, sparse hair present from birth on scalp, arms and legs. Sparing of eyebrows and eyelashes, with no other dysmorphic features, no hearing or neurological abnormalities [12].
Hypotrichosis 10 (HYPT10)	Localised autosomal recessive hypotrichosis 5 (LAH5)	AR	Unknown 7p22.3-p21.3	Complete absence of scalp hair from birth. Sparse eyebrow, eyelash, beard and body hair. Papules present on scalp at the termination of hair follicles [13].

(Continues)

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TABLE 1 | (Continued)

OMIM Name	Disease	Inheritance	Gene and locus	Phenotype
Hypotrichosis 11 (HYPT11)	Hereditary hypotrichosis simplex type 3 (HSS3)	AD	SNRPE 1q32.1	Sparse to complete absence of scalp, eyelash, eyebrow and axillary and body hair from birth. Pubic hairs spared [14, 15].
Hypotrichosis 12 (HYPT12)	Hereditary hypotrichosis simplex type 4 (HSS4)	AD	RPL21 13q12.2	Normal hair at birth and progressive hair loss 2–6 months of age, progressing to sparse or complete loss of scalp hair. Generalised thin, fragile, dry and sparse hair; some with affected eyebrows, eyelashes and body hair. Sparing of beard hair [16, 17].
Hypotrichosis 13 (HYPT13)	Hypotrichosis with woolly hair	AD	KRT71 12q13.13	Tightly curled scalp woolly hair from birth with limited length. No hair shaft or follicular abnormalities. Affecting scalp, eyebrow and eyelash hairs [18].
Hypotrichosis 14 (HYPT14)	Hypotrichosis 14	AR	LSS 21q22.3	Sparse to absent lanugo-like scalp hair, short brittle eyelashes and eyebrows, and sparse body hair [19].
HYPTSV	Hypotrichosis and recurrent skin vesicles	AR	DSC3 18q12.1	Sparse to absent scalp hair; absence of eyebrows, eyelashes, axillary and body hair and cutaneous vesicles of < 1 cm in diameter on scalp and skin [20, 21].
—	Digenic autosomal recessive hypotrichosis	AR	CDH3 16q22.1 & Unknown 12q21.2-q22	Sparse to absent scalp hair from birth, with sparse to normal eyebrows and eyelashes and facial, axillary and body hair [22].

Note: Colours denote broad groups of disease phenotypes.
Abbreviations: AD = autosomal dominant, AR = autosomal recessive [23, 24].

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Table 1: Summarizes the types of hereditary sensory autonomic neuropathy (HSAN)[4-6]

Type	Gene	Inheritance	Locus	Clinical features	Age of onset
HSAN IA (most common)	SPTLC1	AD	9q22.2	Predominant loss of pain and temperature Preservation of vibration sense Lancinating pain Variable distal motor involvement	Adult
HSAN IB	Unknown	AD	3p24-p22	Predominant sensory neuropathy (cough and gastroesophageal reflux) rarely foot ulceration	Adult
HSAN IC	RAB7	AD	3q21	Prominent distal motor loss Sensory loss of all qualities Acro-mutilations	Adult
HSAN II	WNK1/HSN2	AR	12p13.3	Predominant sensory loss Mutilation of hands and feet. Acropathy	childhood
HSAN III	IKBKAP Riley Day dysautonomia	AR	9q31	Prominent autonomic dysfunction Absent fungiform papillae Alacrimia Absent sweating	Congenital
HASN IV	NTRK 1 Congenital Insenstivity to pain	1q21	AR	Reduced pain sensation Anhidrosis Episodic fever Mild Mental retardation Cornea lesions and joint deformities	Congenital
HSAN V	NGFB	1p13.1	AR	Painless fracture Joint deformities Insensitivity to pain Normal intelligence	Congenital
HSAN with spastic paraplegia	CCT5	5p15	AR	Prominent sensory neuropathy Mutilating acropathy Spastic paraplegia	Early childhood

DISORDERS CHARACTERIZED BY DIFFUSE PIGMENTARY DILUTION IN WHICH THE GENETIC DEFECT IS KNOWN			
Disorder	**Gene**	**Protein product**	**Comments**
Oculocutaneous albinism (OCA) **[~40% of patients have OCA1 and ~50% have OCA2]**			
OCA1A	*TYR*	Tyrosinase	• Complete absence of tyrosinase activity and melanin production • Retention of misfolded tyrosinase protein within the ER
OCA1B	*TYR*	Tyrosinase	• Decreased tyrosinase activity; can produce pheomelanin • Variant with temperature-sensitive tyrosinase (normal activity at 35°C, but diminished at 37°C) • Additional variants: minimal pigment, platinum, yellow
OCA2	*OCA2*	P protein (*OCA2* was previously known as *P*)	• Melanosomal transmembrane protein that is also present in the ER • Possible functions include regulating organelle pH, facilitating vacuolar accumulation of glutathione, and processing/trafficking of tyrosinase
OCA3	*TYRP1*	Tyrosinase-related protein 1*	• TYRP1 stabilizes tyrosinase in mice and humans, and it can function as a DHICA oxidase • Both "mutant" TYRP1 and tyrosinase are retained in the ER and then degraded • Rufous phenotype ≫ brown phenotype; latter seen in OCA2
OCA4	*SLC45A2*	Solute carrier family 45 member 2 (previously known as MATP)	• Variable phenotype, with hair ranging from white to yellow–brown; most common in Asians • Transmembrane transporter with a role in tyrosinase processing and intracellular trafficking of proteins to the melanosome
OCA5	Not known	–	• Locus linked to 4q24
OCA6	*SLC24A5*	Solute carrier family 24 member 5	• Cation exchanger that may be involved in ion transport in melanosomes
OCA7	*C10orf11*	Chromosome 10 open reading frame 11	• Expressed in melanoblasts and melanocytes • Possible role in melanocyte differentiation
Hermansky–Pudlak syndrome (HPS)†			
HPS1	*HPS1*	HPS1 (BLOC-3 subunit 1)	• Defective trafficking of organelle-specific proteins to melanosomes, lysosomes and cytoplasmic granules (including platelet dense granules and lytic granules in cytotoxic T lymphocytes) • Pulmonary fibrosis and granulomatous colitis • Component of BLOC-3 (see Fig. 65.8)
HPS2	*AP3B1*	Adaptor related protein complex 3 β1 subunit	• AP-3 recognizes sorting signals within cytosolic tails of cargo molecules and is involved in the trafficking of proteins from the trans-Golgi network to appropriate organelles (see Fig. 65.8) • Pulmonary fibrosis (some patients) • Abnormal targeting of CD1 may play a role in associated immunodeficiency
HPS3	*HPS3*	HPS3 (BLOC-2 subunit 1)	• Component of BLOC-2
HPS4	*HPS4*	HPS4 (BLOC-3 subunit 2)	• Pulmonary fibrosis • Component of BLOC-3
HPS5	*HPS5*	HPS5 (BLOC-2 subunit 2)	• Component of BLOC-2
HPS6	*HPS6*	HPS6 (BLOC-2 subunit 3)	• Component of BLOC-2
HPS7	*DTNBP1*	Dystrobrevin binding protein 1	• Component of BLOC-1
HPS8	*BLOC1S3*	BLOC1S3 (BLOC-1 subunit 3)	• Component of BLOC-1
HPS9	*BLOC1S6*	BLOC1S6 (BLOC-1 subunit 6)	• Component of BLOC-1
HPS10	*AP3D1*	Adaptor related protein complex 3 δ1 subunit	• See AP-3 above • Neurologic impairment (e.g. seizures), immunodeficiency
Chédiak–Higashi syndrome			
	LYST	Lysosomal trafficking regulator	• Abnormal vesicle trafficking and fission/fusion of lysosome-related organelles result in giant organelles (e.g. melanosomes, neutrophil granules [lysosomes], platelet dense granules)
Griscelli syndrome			
GS1	*MYO5A*	Myosin Va (attaches melanosomes to actin filaments)	• In all three forms, melanosomes are retained in the body of the melanocyte rather than trafficking to the tips of dendrites for transfer to keratinocytes (see Fig. 65.10) • Silvery hair seen in all three forms • Myosin Va expressed in neurons (as well as melanocytes) and dysfunction leads to neurologic abnormalities
GS2	*RAB27A*	RAB27A (melanosomal membrane GTPase that binds melanophilin)	• GTPase also expressed in hematopoietic cells; defective release of granule contents from cytotoxic T cells leads to recurrent infections and hemophagocytic syndrome
GS3	*MLPH*	Melanophilin (links myosin Va and RAB27A)	• Melanophilin only expressed in melanocytes, so only pigmentary dilution in type 3
	MYO5A	Myosin Va F-exon deletion	• F-exon only expressed in melanocytes

*Recognized by Mel-5 antibody.
†Reflecting a founder effect, individuals of Puerto Rican origin have HPS1 and HPS3 (3 : 1 ratio); non-Puerto Ricans most commonly have HPS1, followed by HPS3 and HPS4 with ~70% of patients having one of these three types.

Table 65.1 Disorders characterized by diffuse pigmentary dilution in which the genetic defect is known. See Chapter 66 for details of clinical findings. BLOC-1 promotes endosomal maturation by recruiting the Rab5 GTPase-activating protein Msb3; BLOC-2 targets recycling endosomal tubules to melanosomes for cargo delivery; BLOC-3 functions as a Rab32/38 guanine nucleotide exchange factor that is capable of activating small GTPases (activated Rab 32/38 is needed for transport of tyrosinase and TYRP1 to melanosomes). AP-3, adaptor protein complex 3; BLOC, *b*iogenesis of *l*ysosome-related *o*rganelles *c*omplex; DHICA, 5,6-dihydroxyindole-2-carboxylic acid; ER, endoplasmic reticulum; MATP, membrane associated transporter protein; TYRP, tyrosinase-related protein.

and myopia.

Table 49.3: Xeroderma pigmentosum complementation groups. Adapted from Giordano et al. 2016, (Tables 49.2 and 49.3).[183]

Complementation group	*Protein*	*Photosensitivity*	*Cancer predisposition*	*Neurologic abnormalities*
A	XPA	+++	+++	+++
B	XPB and ERCC3	+++	+++	+++
C	XPC	+ to +++	+++	Rare
D	XPD and ERCC2	+ to +++	– to +++	>50%
E	XPE and DDB2	+ to +++	+	Rare
F	XPF and ERCC4	+ to +++	+	+ to +++
G	XPG and ERCC5	+ to +++	+ to +++	– to +++
Variant	Pol η	+++	+++	Rare

(*DDB2*: DNA damage-binding protein 2; *ERCC*: excision repair cross-complementing; *XP*: xeroderma pigmentosum)

CLASSIFICATION SCHEME FOR APLASIA CUTIS CONGENITA (ACC)			
	Inheritance (Gene)	**Features**	**Associated abnormalities**
Group 1: Scalp ACC without multiple anomalies	Sporadic > AD	Usually at the vertex Membranous Hair collar	**No associated abnormalities** **Isolated abnormalities** (most often associated with membranous ACC): cleft lip and/or palate, tracheoesophageal fistula, high myopia and cone-rod dysfunction, patent ductus arteriosus, intestinal lymphangiectasia, omphalocele, polycystic kidneys, double cervix and uterus, intellectual disability, temporal triangular alopecia
	Often AD (*BMS1*)	Usually at the vertex Irregular and scar-like	
Group 2: Scalp ACC associated with Adams–Oliver syndrome (types 1–6)	AD (*ARHGAP31*, *RBPJ*, *NOTCH1, DLL4*) AR (*DOCK6*, *EOGT*)	Midline scalp Often large, irregular lesions Skull defect Dilated scalp veins	**Adams–Oliver syndrome**: terminal transverse limb defects (e.g. reductions, syndactyly, nail dystrophy), cutis marmorata telangiectatica congenita, cardiac malformations, CNS abnormalities
Group 3: Scalp ACC associated with epidermal and organoid nevi (and, occasionally, large congenital melanocytic nevi)	Sporadic due to mosaic mutations (e.g. *HRAS*, *KRAS*)	Scalp, often unilateral Membranous	**Epidermal nevus or nevus sebaceus**, usually in proximity to the ACC **Large congenital melanocytic nevus**: in a few reports* **Ipsilateral ophthalmologic abnormalities**: corneal opacities, scleral dermoids, eyelid colobomas, retinal dystrophy **CNS abnormalities**: intellectual disability, seizures
Group 4: ACC overlying embryologic malformations	Variable	Scalp, often with a hair collar if overlying a neural tube defect Lumbosacral Chest, abdomen	**Cranial malformations**: cephalocele, heterotopic brain or meningeal tissue, congenital midline porencephaly, holoprosencephaly, craniosynostosis, vascular malformation/aplasia/arteriovenous fistula **Spinal malformations**: occult dysraphism, myelomeningocele **Thoracic malformations**: sternal cleft **Abdominal malformations**: omphalocele, gastroschisis **Multiple malformations associated with generalized, lethal ACC ("systemic" ACC)**: corneal opacities, choanal atresia, hypoplastic lungs, absent greater omentum, intestinal malrotation & atresias, hepatosplenomegaly, skull defects, syndactyly, anonychia
Group 5: ACC associated with fetus papyraceus, placental infarcts, or other ischemic events	Sporadic	Scalp, chest, flanks, axillae and/or extremities (see Fig. 64.17) Multiple, symmetric Stellate/angulated configuration	**Associated fetus papyraceus** due to second-trimester death of twin/triplet or fetal reduction of higher-order pregnancies **Placental infarction**, single umbilical artery **Duodenal, ileal, and/or biliary atresias with intestinal infarction** Volkmann ischemic contracture of the forearm Intracranial hemorrhage/CNS abnormalities Amniotic bands, clubbed hands/feet, nail dystrophy
Group 6: ACC associated with EB	AD or AR, depending on EB type	Lower extremities ("Bart syndrome") or large areas of the trunk and extremities	**Simplex, junctional and dystrophic (including bullous dermolysis of the newborn) types of EB**: blistering of skin and/or mucous membranes, skin fragility, nail dystrophy **In more severe cases (most associated with junctional EB)**: pyloric atresia, ureteral stenosis, renal abnormalities, arthrogryposis, abnormal ears and nose, amniotic bands
Group 7: ACC localized to extremities without blistering	AD or AR	Pretibial areas, extensor forearms, dorsal hands and feet	**Radial dysplasia** (associated with ACC on the extensor forearm)
Group 8: ACC caused by specific teratogens, maternal conditions or intrauterine infections	Not inherited	Scalp with drugs Any site with congenital infections	**Methimazole (thiamazole), carbimazole**: imperforate anus, urachal malformations **Misoprostol** **Low-molecular-weight heparin** **Valproic acid** **Maternal antiphospholipid syndrome** **Intrauterine varicella, herpes simplex or rubella infection**: signs of these infections
Group 9: ACC associated with malformation syndromes	Variable	Variable	See Table 64.4 **Amniotic band sequence and disorganization syndrome** (see text)

*Referred to as "SCALP" syndrome (*s*ebaceous nevus, *C*NS malformations, *A*CC, *l*imbal dermoid, *p*igmented nevus).

Table 64.3 Classification scheme for aplasia cutis congenita (ACC). Bold text in column 4 signifies main topics, followed by details. AD, autosomal dominant; AR, autosomal recessive; *ARHGAP31*, Rho GTPase activating protein 31; *BMS1*, ribosome biogenesis factor; CNS, central nervous system; *DLL4*, Delta-like 4; DOCK6, dedicator of cytokinesis 6; EB, epidermolysis bullosa; *EOGT*, EGF domain-specific O-linked N-acetylglucosamine transferase; *RBPJ*, recombination signal binding protein for immunoglobulin κ J region (transcriptional regulator for the Notch pathway). *Adapted from Frieden IJ. J Am Acad Dermatol. 1986;14:646–60, with permission.*

TABLE 12.1. Nomenclature of the parapsoriases.

Disease entity	Synonyms
Large-plaque parapsoriasis[a,b]	Large-plaque (atrophic) parapsoriasis[c]
	LPAP[c]
	Atrophic parapsoriasis[c]
	Poikilodermatous parapsoriasis[c]
	Parapsoriasis en plaques,[c] large-plaque type
	Parapsoriasis en plaques,[d] atrophic type
	Parapsoriasis en grandes plaques simples
	Parapsoriasis en grandes plaques poikilodermiques
	Lichenoid stage of mycosis fungoides
	Poikilodermic mycosis fungoides
	Prereticulotic dermatitis
	Prereticulotic poikiloderma
	Parapsoriasis en plaques[b,d,e]
	Poikiloderma vasculare atrophicans[e,f]
	Parapsoriasis lichenoides[e,g]
Variant: retiform parapsoriasis[h]	Parapsoriasis variegata[c,g]
	Parakeratosis variegata[c]
	Lichen variegatus
	Parapsoriasis lichenoides[e,g]
Small-plaque parapsoriasis[a,b]	Parapsoriasis en plaque,[d] small-plaque type
	Parapsoriasis en plaques,[d] simple discrete type
	Parapsoriasis en plaques,[d] benign type
	Leopard-spot parapsoriasis
	Chronic superficial dermatitis[e]
	Parapsoriasis en plaques[b,d,e]
Variant: digitate dermatosis[i]	Fingerprint parapsoriasis[i]
Variant: xanthoerythroderma perstans[i,j,k]	
Pityriasis lichenoides[l]	Parapsoriasis guttata[c,e,g]
Acute[l]	Pityriasis lichenoides et varioliformis acuta[c]
	PLEVA[c]
	PLVA[c]
	Mucha-Haberman disease[c]
	Parapsoriasis lichenoides et varioliformis acutea
	Parapsoriasis violiformis
	Parapsoriasis acuta[g]
	Acute guttage parapsoriasis
	Acute pityriasis lichenoides
Chronic[l]	Pityriasis lichenoides chronica[c,g]
	Parapsoriasis lichenoides chronica[g]
	Chronic guttate parapsoriasis

Source: Adapted from Lambert, WC, Everett MA: The nosology of parapsoriasis. *J Am Acad Dermatol* 1981; 5:373–395.

[a] Some authorities consider large-plaque and small-plaque parapsoriasis as a single entity, usually termed *parapsoriasis en plaques*.

[b] The term *Brocq's disease* is sometimes utilized to denote small- and large-plaque parapsoriasis together as a single entity; less commonly it is used to denote one or more of the other parapsoriases.

[c] Preferred synonyms, based on frequent usage.

[d] "Parapsoriasis en plaques" is pronounced with the final "s" silent, misleading some English-speaking authors to misspell it "parapsoriasis en plaque."

[e] These terms are frequently used by different authors to denote completely different diseases. For this reason, their use is not recommended.

[f] Poikiloderma vasculare atrophicans is sometimes labeled poikiloderma atrophicans vasculare. Either term is acceptable.

[g] When equivalent terms in different languages are in common use, only the Latin name is given: e.g., parapsoriasis guttata (guttate parapsoriasis, parapsoriasis en gouttes, or en gotas); parapsoriasis lichenoides (lichenoid parapsoriasis, parapsoriasis lichenoides); parapsoriasis variegata (variegate parapsoriasis).

[h] Some authorities regard retiform parapsoriasis as a separate entity.

[i] Some authors regard digitate dermatosis, of which fingerprint parapsoriasis is a synonym and xanthoerythrodermia perstans a variant, as a separate entity.

[j] Xanthoerythrodermia perstans is a variant of digitate dermatosis in which the lesions are yellow in color and on clinical examination. It sometimes incorrectly applied to other forms of parapsoriasis with yellow lesions.

[k] Xanthoerythrodermia perstans is an unnecessary term that should be avoided.

[l] Some authors consider acute and chronic pityriasis lichenoides as separate entities.

TYPES OF SEVERE COMBINED IMMUNODEFICIENCY (SCID)			
Disorder	**Gene**	**Protein function and specific clinical features**	**Cells**
Defects in cytokine receptors and their signaling			
X-linked SCID	*IL2RG*	Common γ chain (γ_c) of the receptors for IL-2, 4, 7, 9, 15, and 21	$T^-/B^+/NK^-$
JAK3 deficiency	*JAK3*	Tyrosine kinase with a role in signal transduction via the receptors for IL-2, 4, 7, 9, 15, and 21	$T^-/B^+/NK^-$
Interleukin-7 receptor deficiency	*IL7R*	α-chain of the IL-7 receptor; critical for development and differentiation of T cells; possible Omenn syndrome phenotype	$T^-/B^+/NK^+$
Increased apoptosis of lymphocytes ± other hematopoietic cells			
Defects in purine pathway enzymes that result in the build-up of toxic metabolites			
Adenosine deaminase (ADA) deficiency	*ADA*	Enzyme in the purine salvage pathway	$T^-/B^-/NK^-$
Purine nucleoside phosphorylase (PNP) deficiency	*PNP*	Enzyme in the purine salvage pathway (downstream of ADA)	$T^-/B^-/NK^-$
Defects disrupting mitochondrial energy production			
Reticular dysgenesis (aleukocytosis)	*AK2*	Adenylate kinase 2, a mitochondrial enzyme that regulates ADP levels in leukocytes; profound neutropenia and sensorineural deafness	$T^-/B^-/NK^-$
Defects disrupting actin polymerization			
Coronin 1A deficiency	*CORO1A*	Actin regulator expressed in T-cell precursors; severe warts, molluscum contagiosum, and herpesvirus infections	$T^-/B^+/NK^+$
Defects in (pre-)T-cell receptors (TCRs) ± B-cell receptors (BCRs) and their signaling			
Omenn syndrome	*RAG1* and *RAG2*	Mediate V(D)J recombination of the genes that encode BCRs (immunoglobulins) and TCRs§; erythroderma, alopecia, LAN, HSM, diarrhea, eosinophilia, elevated IgE levels (see text)	$T^-/B^-/NK^+$
Artemis deficiency	*DCLRE1C (Artemis)*	Repairs the DNA double-stranded breaks that occur during V(D)J recombination and from ionizing radiation§; possible Omenn syndrome phenotype	$T^-/B^-/NK^+$
PRKDC deficiency	*PRKDC*	DNA-activated protein kinase catalytic subunit critical for V(D)J recombination§	$T^-/B^-/NK^+$
LIG4 syndrome	*LIG4*	Ligase component of the DNA non-homologous end-joining (NHEJ) machinery required for the repair of DNA double-stranded breaks§; photosensitivity, telangiectasias, bird-like facies, microcephaly	$T^-/B^-/NK^+$
NHEJ1 (Cernunnos) deficiency	*NHEJ1*	NHEJ factor required for the repair of DNA double-stranded breaks§; bird-like facies, microcephaly	$T^-/B^-/NK^+$
T-cell receptor deficiencies	*CD3γ, CD3δ, CD3ε,* and *CD3ζ*	TCR-associated proteins required for cell surface expression of and signal transduction by the TCR; most severe with defective CD3δ	$T^-/B^+/NK^+$
	TRAC	TCR-α constant region; relatively mild immunodeficiency	
CD45 deficiency	*PTPRC*	Transmembrane tyrosine phosphatase with a role in TCR signaling	$T^-/B^+/NK^+$
ZAP70 deficiency	*ZAP70*	Tyrosine kinase with a role in TCR signaling	$CD4^+CD8^-/B^+/NK^+$
CD8 deficiency¶	*CD8A*	Co-receptor for TCRs recognizing MHC class I-associated peptides; role in TCR signaling	$CD4^+CD8^-/B^+/NK^+$
Calcium channel deficiency	*ORAI1*	Pore-forming subunit of calcium channel with a role in TCR signaling; hypohidrotic ectodermal dysplasia and myopathy	$T^+/B^+/NK^+$
	STIM1	Calcium channel component with a role in TCR signaling; LAN, HSM, autoimmune cytopenias, dental dysplasia, myopathy	$T^+/B^+/NK^+$
Antigen presentation defects (bare lymphocyte syndromes)			
MHC class II deficiency	*CIITA, RFXANK, RFX5, and RFXAP*	Transactivate the MHC class II promoter; MHC class II molecules are required for the positive selection of $CD4^+$ T cells in the thymus as well as for antigen presentation to $CD4^+$T cells	$T^+/B^+/NK^+$, but fewer and dysfunctional $CD4^+$ T cells
MHC class I deficiency¶	*TAP1*, *TAP2, TAPBP, B2M*	*T*ransporter associated with *a*ntigen *p*rocessing (TAP) and the TAP binding protein (tapasin); translocate peptides from the cytosol to MHC class I molecules in the ER; cutaneous granulomas in TAP deficiency	$CD4^+CD8^-/B^+/NK^+$
Defective thymic development			
Winged helix nude (forkhead box N1) deficiency	*WHN (FOXN1)*	Transcription factor important to thymic development; complete congenital alopecia and nail dystrophy (human equivalent of "nude" mice)	$T^-/B^+/NK^+$

§Patients have increased sensitivity to ionizing radiation.
¶Not true SCID

Table 60.16 Types of severe combined immunodeficiency (SCID). "Leaky SCID" refers to less severe deficits in T-cell number and function due to hypomorphic (milder) mutations in SCID-associated genes. Cartilage–hair hypoplasia (see text) also occasionally presents as SCID with an Omenn-like phenotype. Two most common forms of SCID, accounting for approximately two-thirds of cases, are shaded. B2M, β2-microglobulin; *DCLRE1C*, DNA cross-link repair protein 1C; ER, endoplasmic reticulum; HSM, hepatosplenomegaly; LAN, lymphadenopathy; MHC, major histocompatibility complex; V(D)J, variable/diversity/joining.

defective function of both cell-mediated and humoral immunity [1–3]. The combined frequency of all forms of SCID is 1 in 30,000 to 1 in 100,000 live births, and approximately 75% of patients are male. Almost half of affected individuals have an X-linked recessive form of SCID, which is usually caused by defects in the gene encoding the common γ (γ_c) chain of the IL-12 receptor. Other forms of SCID have autosomal recessive inheritance and include adenosine deaminase (ADA) deficiency (20% of SCID) and JAK3 deficiency (approximately 5% of SCID).

Pathogenesis. The molecular defects that can lead to a SCID phenotype are summarized in Table 177.8 [1,2].

Clinical features. Patients with SCID typically begin to have recurrent infections, chronic diarrhoea and failure to thrive during the first few months of life. Early infections often include mucocutaneous candidiasis, persistent viral gastroenteritis and pneumonias resulting from bacteria, viruses or *P. jiroveci*. Skin infections are commonly caused by *S. aureus* and *Strep. pyogenes* as well as *Candida* spp. Despite their frequent infections, patients with SCID usually lack tonsillar buds and palpable lymphoid tissue.

Infants with SCID may present with morbilliform or seborrhoeic dermatitis-like eruptions [4,5], which often reflect maternofetal GVHD (see Chapter 178). Cutaneous GVHD in patients with SCID may also resemble lichen planus, acrodermatitis enteropathica, Langerhans' cell histiocytosis, ichthyosiform erythroderma or scleroderma. Skin biopsy specimens show the histopathological characteristics of GVHD. Extensive eczematous dermatitis or severe exfoliative erythroderma, often associated with alopecia, may occur without GVHD in infants with Omenn syndrome. Additional features of Omenn syndrome include lymphadenopathy, hepatosplenomegaly and peripheral eosinophilia.

Most patients with SCID have a profound deficiency of T cells and a low absolute lymphocyte count. Patients are further classified by the results of fluorescent-activated cell sorter analysis into those with and without B and NK cells. Diagnosis of a particular form of SCID can be established by genetic testing, flow cytometric analysis of peripheral blood mononuclear cells with antibodies directed against a particular protein (e.g. γ_c or JAK3) that is missing from the cell surface, or measurement of red blood cell levels of a defective enzyme (e.g. ADA) or purine metabolites. In families with a previously affected sibling, prenatal diagnosis of SCID has been performed by fetal DNA analysis and ADA assays.

Maternal T-cell engraftment is detectable (requiring maternal cells to comprise ≥1% of peripheral blood leucocytes) in approximately 50% of infants with SCID [5]. In the majority of cases, the presence of these maternal T cells is asymptomatic; in 30–40% of patients, mild findings such as morbilliform eruptions, cutaneous erythema

Table 177.8 Severe combined immunodeficiency (SCID)

Disorder	Gene	Protein function ± specific clinical findings	Cells
Defects in cytokine receptors and their signalling			
X-linked SCID	*IL2RG*	Common γ chain (γ_c) of the receptors for IL-2, 4, 7, 9, 15 and 21	$T^-/B^+/NK^-$
JAK3 deficiency	*JAK3*	Tyrosine kinase with a role in signal transduction via the receptors for IL-2, 4, 7, 9, 15 and 21	$T^-/B^+/NK^-$
Interleukin-7 receptor deficiency	*IL7R*	α chain of the IL-7 receptor; critical for development and differentiation of T cells; occasionally Omenn-like phenotype	$T^-/B^+/NK^+$
Increased apoptosis of lymphocytes ± other haematopoietic cells			
Accumulation of toxic metabolites due to purine pathway enzyme defects			
ADA deficiency	*ADA*	Enzyme in the purine salvage pathway; cupping/flaring of the costochondral junction on chest X-rays; occasionally Omenn-like phenotype	$T^-/B^-/NK^-$
PNP deficiency	*PNP*	Enzyme in the purine salvage pathway that acts downstream of ADA	$T^-/B^-/NK^-$
Defects disrupting mitochondrial energy production			
Reticular dysgenesis (aleucocytosis)	*AK2*	Adenylate kinase 2, a mitochondrial enzyme that regulates ADP levels in leucocytes; profound neutropenia and sensorineural deafness	$T^-/B^-/NK^-$
Defects disrupting actin polymerization			
Coronin 1A deficiency	*CORO1A*	Actin regulator expressed in T-cell precursors	$T^-/B^+/NK^+$

Table 177.8 *Continued*

Disorder	Gene	Protein function ± specific clinical findings	Cells
Defects in (pre-) TCRs +/– BCRs and their signalling			
Omenn syndrome	*RAG1* and *RAG2*	Mediate V(D)J recombination of genes that encode BCRs (immunoglobulins) and TCRs; exfoliative erythroderma, hepatosplenomegaly, lymphadenopathy, eosinophilia	$T^-/B^-/NK^+$
Artemis deficiency	*DCLRE1C (Artemis)*	DNA cross-link repair protein that repairs DNA double-stranded breaks that occur during V(D)J recombination and upon exposure to ionizing radiation; sensitivity to ionizing radiation; occasionally Omenn-like phenotype	$T^-/B^-/NK^+$
PRKDC deficiency	*PRKDC*	DNA-activated protein kinase catalytic subunit critical for V(D)J recombination; sensitivity to ionizing radiation	$T^-/B^-/NK^+$
LIG4 syndrome	*LIG4*	Ligase component of DNA non-homologous end-joining machinery required for repair of DNA double-stranded breaks; sensitivity to ionizing radiation; variable severity, occasionally Omenn-like phenotype; microcephaly, bird-like facies, photosensitivity and telangiectasias	$T^-/B^-/NK^+$
NHEJ1 (Cernunnos) syndrome	*NHEJ1*	Non-homologous end-joining factor required for the repair of DNA double-stranded breaks; sensitivity to ionizing radiation; variable severity; microcephaly and bird-like facies	$T^-/B^-/NK^+$
TCR deficiencies	CD3γ, CD3δ, CD3ε and CD3ζ	TCR-associated proteins required for cell-surface expression of and signal transduction by the TCR; the T-cell defect is most severe with CD3δ defects	$T^-/B^+/NK^+$
CD45 deficiency	*PTPRC*	Transmembrane tyrosine phosphatase with a role in TCR signalling	$T^-/B^+/NK^+$
ZAP-70 deficiency	*ZAP-70*	Tyrosine kinase with a role in TCR signalling; lymphocytosis; variable severity and phenotype, occasionally Omenn-like	$CD4^+CD8^-/B^+/NK^+$
CD8 deficiency	*CD8A*	Co-receptor for TCRs recognizing MHC class I-associated peptides; role in TCR signalling; milder than true SCID	$CD4^+CD8^-/B^+/NK^+$
Calcium channel deficiency	*ORAI1*	Pore-forming subunit of calcium channel with a role in TCR signalling; hypohidrotic ectodermal dysplasia and myopathy	$T^+/B^+/NK^+$
	STIM1	Calcium channel component with a role in TCR signalling; hepatosplenomegaly, lymphadenopathy, autoimmune cytopenias, dental dysplasia, myopathy	$T^+/B^+/NK^+$
Antigen presentation defects (bare lymphocyte syndromes)			
MHC class II deficiency	*CIITA* *RFXANK* *RFX5* *RFXAP*	Transactivate the MHC class II promoter; MHC class II molecules are required for the positive selection of $CD4^+$ T cells in the thymus as well as for antigen presentation to $CD4^+$ T cells	$T^+/B^+/NK^+$; fewer/ dysfunctional $CD4^+$ T cells
MHC class I deficiency	*TAP1, TAP2* and *TAPBP*	TAP and TAP binding protein (tapasin); translocate peptides from the cytosol to MHC class I molecules in the endoplasmic reticulum; milder than true SCID; cutaneous granulomas	$CD4^+CD8^-/B^+/NK^+$
Defective thymic development			
Winged helix nude (forkhead box N1) deficiency	*WHN (FOXN1)*	Transcription factor important to thymic development; congenital alopecia and nail dystrophy (human equivalent of 'nude' mice)	$T^-/B^+/NK^+$

Cartilage–hair hypoplasia (see text) occasionally presents as SCID with an Omenn-like phenotype.
ADA, adenosine deaminase; BCR, B-cell receptor; IL, interleukin; MHC, major histocompatibility complex; NK, natural killer; PNP, purine nucleoside phosphorylase; TAP, transporter associated with antigen processing; TCR, T-cell receptor; V(D)J, variable/diversity/joining.
Adapted with permission from Bolognia JL, Jorizzo JL, Rapini RP, eds. *Dermatology*, 2nd edn. London: Elsevier, 2007.

TABLE I. Ectodermal Dysplasias of Group A (N = 163), Modified From Visinoni et al. [2009]

	Ectodermal dysplasia (ED)	Reference (OMIM, whenever available)	Inheritance
	Subgroup hair–teeth–nails–sweat glands. N = 38		
1	Alopecia-contractures-dwarfism mental retardation syndrome	203550	AR
2	Ankyloblepharon-ectodermal defects-cleft lip/palate syndrome (AEC; Hay–Wells syndrome); **Rapp–Hodgkin syndrome (129400) included**	106260	AD
3	Anonychia with flexural pigmentation	106750	AD
4	AREDYLD (Acrorenal field defect, ED, and lipoatrophic diabetes)	207780	AR
5	Arthrogryposis and ED	601701	AR
6	Camarena syndrome	Freire-Maia and Pinheiro [1984]	AD?; XD?
7	Cleft lip/palate-ED syndrome (CLPED1; Zlotogora–Ogur syndrome; Margarita Island syndrome)	225060	AR
8	Curly hair-acral keratoderma-caries syndrome	van Steensel and van der Hout [2009]	AD
9	Dyskeratosis congenita, AD (Dyskeratosis congenita, Scoggins type)	127550	AD
10	Dyskeratosis congenita, AR	224230	AR
11	Dyskeratosis congenita, X-linked (Zinsser–Cole–Engman syndrome)	305000	XR
12	Ectrodactyly, ED, and cleft lip/palate syndrome (EEC1)	129900	AD
13	Ectrodactyly, ED, and cleft lip/palate syndrome 3 (EEC3)	604292	AD
14	ED hypohidrotic, with acanthosis nigricans (Lelis syndrome)	608290	?
15	**ED-syndactyly syndrome 2 (EDSS2)**	613576	AR
16	ED with natal teeth, Turnpenny type	601345	AD
17	ED, Caratinga type	Montebelo et al. [1996]	AD?; XD?
18	ED, hypohidrotic, with hypothyroidism and agenesis of the corpus callosum	225040	AD?; AR?; XD?
19	Focal dermal hypoplasia (FDH)	305600	XD
20	Hypohidrotic ED, autosomal dominant (ADHED)	129490	AD
21	Hypohidrotic ED, autosomal recessive (ARHED)	224900	AR
22	Hypohidrotic ED, X-linked (XLHED; Christ–Siemens–Touraine syndrome; CST syndrome)	305100	XR
23	Hypohidrotic ED with immune deficiency	300291	XD
24	Hypohidrotic ED with immunodeficiency, osteopetrosis, and lymphedema (OLEDAID syndrome)	300301	XD
25	Hypomelanosis of Ito (HMI, Incontinentia pigmenti type I; IP1)	300337	XD
26	Keratitis-ichthyosis-deafness syndrome, autosomal dominant (KID syndrome, AD)	148210	AD
27	Keratitis-ichthyosis-deafness syndrome, autosomal recessive (KID syndrome, AR)	242150	AR
28	Naegeli syndrome (Naegeli–Franceschetti–Jadassohn syndrome, NFJS)	161000	AD
29	Odontoonychodermal dysplasia (OODD); **Schöpf–Schulz–Passarge syndrome (224750) included**	257980	AR
30	Odontotrichomelic syndrome (tetramelic deficiencies, ectodermal dysplasia, deformed ears, and other abnormalities)	273400	AR
31	Pachyonychia congenita, type 1 (PC1)	167200	AD
32	Pachyonychia congenita, type 2 (PC2)	167210	AD
33	Papillon–Lefevre syndrome	245000	AR
34	Rosselli–Gulienetti syndrome	225000	AR
35	Scalp-ear-nipple syndrome (Finlay–Marks syndrome; ED with adrenal cyst)	181270; 129550	AD
36	Tricho-odonto-onychodysplasia with pili torti	Freire-Maia and Pinheiro [1984]	AD?; XD?
37	Tricho-onycho-dental dysplasia (TOD)	Koshiba et al. [1978]	AD
38	Xeroderma-talipes-enamel defects (XTE)	Moynahan [1970]	AR
	Subgroup hair–teeth–nails. N = 34		
39	Ackerman syndrome	200970	AR
40	ADULT syndrome	103285	AD
41	Arthrogryposis, ED, cleft lip/palate, and developmental delay	301815	XR
42	Cardiofaciocutaneous syndrome (CFC)	115150	AD
43	**Cardiomyopathy, dilated, with woolly hair, and keratoderma (Carvajal syndrome)**	605676	AR
44	Clouston syndrome (ectodermal dysplasia, hidrotic, autosomal dominant)	129500	AD

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TABLE I. (*Continued*)

	Ectodermal dysplasia (ED)	Reference (OMIM, whenever available)	Inheritance
45	Coffin–Siris syndrome	135900	AD?; AR?; XD?
46	Costello syndrome	218040	AR
47	**Cranioectodermal dysplasia (CED1, Sensenbrenner syndrome); CED2 (613610), CED3 (614099) and CED4 (614378) included**	218330	AR
48	Dermoodontodysplasia	125640	AD
49	Dolichocephaly, dental defects, trichodysplasia	Freire-Maia and Pinheiro [1984]	AD
50	**ED-syndactyly syndrome 1 (EDSS1); ED with pillous anomaly and syndactyly (**Wiedemann et al., 1978**) included**	613573	AR
51	ED, trichoodontoonychial type	129510	AD
52	Ellis–van Creveld syndrome (EVC)	225500	AR
53	GAPO syndrome (growth retardation, alopecia, pseudoanodontia, and optic atrophy)	230740	AR
54	GOMBO syndrome	233270	AR
55	Hidrotic ED autosomal recessive (Fried's tooth and nail syndrome)	602401	AR
56	Hypotrichosis with pili bifurcate	Beemer et al. [1987f]	AR?
57	Incontinentia pigmenti (IP2)	308300	XD
58	Oculotrichodysplasia (OTD)	257960	AR
59	Odonto-onychodysplasia-alopecia	Pinheiro and Freira Maia [1981]	AR
60	Odontotrichoungual-digital-palmar syndrome	601957	AD?; XD?
61	Pineal hyperplasia, insulin-resistant diabetes mellitus, and somatic abnormalities	262190	AR
62	Rothmund–Thomson syndrome (RTS)	268400	AR
63	Schinzel–Giedion midface-retraction syndrome	269150	AR?; AD?
64	Sener syndrome	606156	?
65	**Split-hand/foot malformation (SHFM1); SHFM3 (246560), SHFM4 (605289), and SHFM5 (606708) included**	183600	AD
66	Thumb deformity and alopecia	188150	AD
67	Trichodentoosseus syndrome (TDO)	190320	AD
68	Tricho-dermodysplasia-dental defects	Pinheiro et al. [1986]	AD?; XD?
69	Trichoodontoonychial dysplasia with bone deficiency	275450	AR?
70	Trichorhinophalangeal syndrome, type I (TRPS1)	190350	AD
71	Trichothiodystrophy, photosensitive (TTDP)	601675	AR
72	Witkop syndrome	189500	AD
	Subgroup hair–teeth–sweat glands. N = 8		
73	Böök syndrome	112300	AD
74	Cleft lip/palate, ED, acral anomalies	Richieri-Costa et al. [1992]	AR
75	Hypohidrotic ED with focal sweating	Gorlin [1988]	AR?; XR?
76	IFAP syndrome with or without bresheck syndrome (Ichthyosis follicularis, atrichia, and photophobia syndrome)	308205	XR
77	Johnson neuroectodermal syndrome	147770	AD
78	Lenz–Passarge dysplasia	Lenz [1963]	XD
79	Leukomelanoderma, infantilism, mental retardation, hypodontia, hypotrichosis	246500	AR
80	Ulnar-mammary syndrome (UMS)	181450	AD
	Subgroup hair–nails–sweat glands. N = 4		
81	Alopecia-skin atrophy-anonychia-tongue defects	Sequeiros and Sack [1985]	?
82	ED, hypohidrotic, with hypothyroidism and ciliary dyskinesia (HEDH syndrome)	225050	AR
83	ED/skin fragility syndrome	604536	AR
84	Fischer–Volavsek syndrome	Fischer [1921]	AD
	Subgroup teeth–nails–sweat glands. N = 2		
85	Ameloonychohypohidrotic syndrome	104570	AD
86	Limb-mammary syndrome (LMS)	603543	AD
	Subgroup hair–teeth. N = 29		
87	Barber–Say syndrome	209885	AR?; AD?; XD?
88	Blepharocheilodontic syndrome	119580	AD
89	Brachymetapody-anodontia-hypotrichosis-albinoidism	211370	AR
90	CAHMR syndrome (Cataract, hypertrichosis, mental retardation syndrome)	211770	AR
91	Cerebellar ataxia and ED	212835	AR

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TABLE I. (*Continued*)

	Ectodermal dysplasia (ED)	Reference (OMIM, whenever available)	Inheritance
92	Cleft lip/palate-oligodontia-syndactyly-hair defects	Martínez et al. [1987]	AD?; XD?
93	Coloboma, congenital heart disease, ichthyosiform dermatosis, mental retardation, and ear anomalies syndrome (CHIME, Zunich neuroectodermal syndrome)	280000	AR
94	Congenital atrichia, palmoplantar hyperkeratosis, mental retardation, and early loss of teeth	Steijlen et al. [1994]	AR?
95	Dubowitz syndrome	223370	AR
96	ED and neurosensory deafness	224800	AR
97	ED, Cape Verde	Werninghaus [1993]	AR
98	EEM syndrome (ED, ectrodactyly, and macular dystrophy); **Hypotrichosis, congenital, with juvenile macular dystrophy (HJMD, 601553) included**	225280	AR
99	Gorlin–Chaudhry–Moss syndrome	233500	AR
100	Hallermann–Streiff syndrome (HSS)	234100	AR
101	Hypertrichosis terminalis, generalized, with our without gingival hyperplasia (Gingival fibromatosis with hypertrichosis)	135400	AD
102	Hypertrichosis universalis	145700	AD
103	Johanson–Blizzard syndrome (JBS)	243800	AR
104	Mental retardation, hypotrichosis, and syndactyly	Lopes and Marques-de-Faria [1996]	AR?
105	Oculodentoosseous dysplasia, recessive	257850	AR
106	Oculodentodigital dysplasia (ODDD)	164200	AD
107	Orofaciodigital syndrome I (OFD1)	311200	XD
108	Pili torti, early onset	261900	AR
109	Pilodental dysplasia with refractive errors	262020	AR
110	Progeroid short stature with pigmented nevi (Mulvihill–Smith syndrome)	176690	AD
111	Rodrigues blindness (Microphthalmia, microcornea, and sclerocornea with short stature and hair and dental abnormalities)	268320	AR
112	Trichodental dysplasia	601453	AD
113	Trichodysplasia and amelogenesis imperfect	Angelos and Jorgenson [1993]	AD?; XD?
114	Uncombable hair, retinal pigmentary dystrophy, dental anomalies, and brachydactyly	191482	AD
115	Walbaum–Dehaene–Schlemmer syndrome	Walbaum et al. [1971]	AR
	Subgroup hair–nails. N = 25		
116	Anonychia-onychodystrophy with hypoplasia or absence of distal phalanges (Cooks syndrome)	106995	AD
117	Autosomal recessive neurodegenerative disorder with trichorrhexis invaginata and ED	Gyure et al. [1992]	AR?
118	Cartilage-hair hypoplasia (CHH)	250250	AR
119	Curly hair-ankyloblepharon-nail dysplasia syndrome (CHANDS)	214350	AR
120	ED hidrotic, Christianson–Fourie type	601375	AD
121	ED with skin anomalies and mental retardation	Halal et al. [1991]	AR
122	ED, "pure" hair-nail type	602032	AD?
123	Hair-nail dysplasia	Pinheiro and Freire-Maia [1992]	AD
124	Hairy elbows (hypertrichosis cubiti)	139600	AD
125	Ichthyosis and male hypogonadism	308200	XR?
126	Ichthyosis with alopecia, eclabion, ectropion, and mental retardation	242510	AR
127	Lymphedema-hypoparathyroidism syndrome	247410	AR?; XR?
128	Monilethrix	158000	AD
129	Onychotrichodysplasia and neutropenia	258360	AR
130	Palmoplantar keratoderma and congenital alopecia, autosomal dominant (alopecia congenita with keratosis palmoplantaris)	104100	AD
131	Pili torti and onychodysplasia	Beare [1952]	AD
132	Pili torti, alopecia and onychodysplasia	Calzavara-Pinton et al. [1991]	AR
133	Polyposis, skin pigmentation, alopecia, and fingernail changes	175500	?
134	Popliteal pterygium syndrome, lethal type	263650	AR
135	**Short stature, onychodysplasia, facial dysmorphism, and hypotrichosis syndrome (SOFT syndrome)**	Sarig et al. [2012]	AR
136	Syndrome of accelerated skeletal maturation, failure to thrive and peculiar face (Marshall syndrome II)	Marshall et al. [1971]	AR?; XR?

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TABLE I. (*Continued*)

	Ectodermal dysplasia (ED)	Reference (OMIM, whenever available)	Inheritance
137	T-cell immunodeficiency, congenital alopecia, and nail dystrophy	601705	AR?
138	Trichomegaly with mental retardation, dwarfism, and pigmentary degeneration of retina	275400	AR
139	Tricho-onychodysplasia-xeroderma	Freire-Maia et al. [1985]	AR
140	Trichothiodystrophy, nonphotosensitive 1 (TTDN1)	234050	AR
	Subgroup hair–sweat glands. N = 4		
141	Dry skin and extranumerary areolae	Freire-Maia and Chautard-Freire-Maia [1990]	AD
142	Focal facial dermal dysplasia (Brauer syndrome); Facial ED (Setleis syndrome, 227260) included	136500	AD
143	Short stature-kidney insufficiency-ophthalmological anomaly-growth retardation-ED (SKORED)	Greenstein et al. [1985]	AR?; XR?
144	Tricho-facio-hypohidrotic syndrome	Antley et al. [1976]	AR?; XR?
	Subgroup teeth–nails. N = 14		
145	Corneodermatoosseous syndrome (CDO syndrome)	122440	AD
146	Deafness, congenital, and onychodystrophy, autosomal dominant	124480	AD
147	Deafness, onychodystrophy, osteodystrophy, and mental retardation syndrome (DOOR syndrome)	220500	AR?; AD?
148	Dermatoosteolysis, Kirghizian type	221810	AR
149	Haim–Munk syndrome (HMS)	245010	AR
150	Hearing loss, sensorineural, with enamel hypoplasia and nail defects (Heimler syndrome)	234580	AR
151	**Khan et al. chondroectodermal dysplasia**	Khan et al. [2012]	AR
152	Lacrimoauriculodentodigital syndrome (LADD)	149730	AD
153	Odontomicronychial dysplasia	601319	AR
154	Odonto-ungueal dysplasia	Pinheiro and Freire-Maia [1996]	AD
155	Otopalatodigital syndrome, type I (OPD1)	311300	XD
156	Pycnodysostosis	265800	AR
157	Weyers acrofacial dysostosis	193530	AD
158	Williams–Beuren syndrome (WBS)	194050	AD
	Subgroup teeth–sweat glands. N = 3		
159	Hypohidrotic ED with mydriasis, iris atrophy, and mental retardation	Beyer et al. [1979]	AD?
160	Kohlschutter–Tonz syndrome (epilepsy, dementia, and amelogenesis imperfecta)	226750	AR?; XR?
161	Marshall syndrome I	154780	AD
	Subgroup nail–sweat glands. N = 2		
162	Adermatoglyphia with congenital facial milia and acral blisters, digital contractures, and nail abnormalities (ED, absent dermatoglyphic pattern, changes in nails, and simian crease)	129200	AD
163	Pachyonychia congenita, autosomal recessive	260130	AR

Note: the bold text highlights information not listed in Visinoni et al. [2009].
AR, autosomal recessive; AD, autosomal dominant; XD, X-linked dominant; XR, X-linked recessive; ?, unknown.

we have consulted the OMIM, PUBMED, and Science Direct online databases.

RESULTS AND DISCUSSION

After the first International Conference on Ectodermal Dysplasias Classification, held in Charleston (South Carolina) in 2008, we listed 186 disorders in an article published in a special issue of the *American Journal of Medical Genetics* [Visinoni et al., 2009]. Within this reported series of ectodermal dysplasias, descriptions of 26 single cases were included because of their significant frequency of conditions (13.8%), and also because that by mentioning these cases the description of similar patients could be encouraged, which could possibly help clarify some poorly understood clinical presentations.

Reviewing the data and following what was decided at the 2012 International Conference on Ectodermal Dysplasias Classification (Charleston, SC), we removed single cases from the list of ectodermal dysplasias.

Table I provides a list of 163 dysplasias belonging to Group A. Besides the exclusion of the single cases and the inclusion of new EDs, new lumpings of conditions were added to this table, and the syndrome nomenclature was reviewed. For 78.5% of these conditions there is an OMIM entry, providing easy access to the more relevant information. The pattern of inheritance is also shown in Table I. It is remarkable that 39.9% are autosomal recessive, 33.7% are autosomal dominant, and 7.4% are X linked disorders. It is also noteworthy that the pattern of inheritance is not established in 19% of the ectodermal dysplasias.

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TABLE II. Ectodermal Dysplasias (N = 7) Not Included in Visinoni et al. [2009]

ED	Reference	Inheritance	Hair	Teeth	Nails	Sweat glands	Other signs
Subgroup hair–teeth–nails–sweat glands							
Cardiomyopathy, dilated, with woolly hair and keratoderma (Carvajal syndrome)	605676	AR	Woolly, wavy, wiry and auburn hair; sparse eyebrows and eyelashes	Hypodontia; diminutive upper lateral incisors; prepubertal periodontitis; premature root resorption of primary teeth	Small and thickened	Hypohidrosis	Dilated cardiomyopathy leading to congestive heart failure; epidermolytic palmoplantar keratoderma
Subgroup hair–teeth–nails							
Cranioectodermal dysplasia (CED1, Sensenbrenner syndrome); CED2 (613610), CED3 (614099) and CED4 (614378) included	218330	AR	Sparse; fine and slow-growing hair	Hypodontia; microdontia; abnormally shaped	Short and broad	Normal	Craniosynostosis; microcephaly; hypoplasic posterior corpus callosum; postnatal growth retardation; narrow thorax with pectus excavatum; short limbs; brachydactyly; nephropathy; photophobia
ED-syndactyly syndrome 1 (EDSS1); ED with pillous anomaly and syndactyly [Wiedemann et al., 1978] included	613573	AR	Sparse, coarse and breakable scalp and body hair, eyebrows and eyelashes; progressive hair loss; alopecia; pilli torti	Enamel hypoplasia; peg-shaped; widely spaced	Hypoplastic	Normal	Cutaneous syndactyly; palmar hyperkeratosis
ED-syndactyly syndrome 2 (EDSS2)	613576	AR	Sparse and slow growing scalp hair; sparse eyebrows and eyelashes, sparse to absent armpit and pubic hair; thin to absent body hair; follicular hyperkeratosis	Enamel hypoplasia; conical; widely spaced	Hypoplastic; thickened, flat, and yellowish or discolored	Hyperhidrosis (hands, face, and scalp)	Cutaneous syndactyly; palmoplantar keratoderma; hard scaly skin; pointed nose, thin upper lips, and large prominent ear pinnae; cardiomegaly
Split-hand/foot malformation (SHFM1); SHFM3 (246560), SHFM4 (605289) and SHFM5 (606708) included	183600	AD	Sparse; alopecia	Hypodontia	Dysplasic	Normal	Mental retardation; ectrodactyly; cleft lip/palate; micrognathia; occasional deafness and cardiac defects
Subgroup hair–nails							
Short stature, onychodysplasia, facial dysmorphism, and hypotrichosis syndrome (SOFT syndrome)	Sarig et al. [2012]	AR	Hypotrichosis; postpubertal sparse and short hair	Normal	Hypoplastic	Normal	Short stature; short and thick long bones; hypoplastic pelvis and sacrum; clinodactyly and brachydactyly; hypoplastic distal phalanges; peculiar faces; small ears; high-pitched voice
Subgroup teeth–nails							
Khan et al. chondroectodermal dysplasia	Khan et al. [2012]	AR	Normal	Absence of mandibular central incisors; misshaped; proeminent	Dysplastic; enlarged nail bed; hypertrophic and convex shaped	Normal	Short stature; osteopenia;with breakable bonés; night blindness; watering eyes; ear infections

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TABLE III. Genes (N = 13[a]) and Chromosomal Regions (N = 6) Responsible for 15 Different Ectodermal Dysplasias Not Included in Visinoni et al. [2009]

Chromossome	Gene	Protein or gene product	Ectodermal dysplasia (ED)	OMIM/Refs
Xp22.12–p22.11	*MBTPS2*	Membrane-bound transcription factor site-2 protease	IFAP syndrome with or without bresheck syndrome	308205
1q23.3	*PVRL4*	Poliovirus receptor-related protein 4	ED-syndactyly syndrome 1 (EDSS1)	613573
2p24.1	*WDR35*	WD repeat-containing protein 35	Cranioectodermal dysplasia (CED1–4)	218330, 613610, 614099, 614378
2q24.1–q31.1			Khan et al. chondroectodermal dysplasia	Khan et al. [2012]
2q31			SHFM1, 3–5 syndrome	183600, 246560, 605289, 606708
2q37.3	*TWIST2*	Twist-related protein 2	Focal facial dermal dysplasia (facial ED)	136500, 227260
3p21.2	*POC1A*	Centriolar protein homolog A	Short stature, onychodysplasia, facial dysmorphism, and hypotrichosis syndrome (SOFT)	Sarig et al. [2012]
3q21.3–q22.1	*IFT122*	Intraflagellar transport protein 122 homolog	Cranioectodermal dysplasia (CED1–4)	218330, 613610, 614099, 614378
4p14	*WDR19*	WD repeat-containing protein 19	Cranioectodermal dysplasia (CED1–4)	218330, 613610, 614099, 614378
6p24.3	*DSP*	Desmoplakin	Cardiomyopathy, dilated, with woolly hair and keratoderma (Carvajal syndrome)	605676
6q22.31	*GJA1*[a]	Connexin 43	Oculodentodigital dysplasia (ODDD)	164200
			Hallermann–Streiff syndrome (HSS)	234100
7p21.2–p14.3			ED syndactyly syndrome 2 (EDSS2)	613576
7q21.2–q21.3			SHFM1, 3–5 syndrome	183600, 246560, 605289, 606708
10q24			SHFM1, 3–5 syndrome	183600, 246560, 605289, 606708
14q24.3	*IFT43*	Intraflagellar transport protein 43 homolog	Cranioectodermal dysplasia (CED1–4)	218330, 613610, 614099, 614378
16p13.3	*ROGDI*	Protein rogdi homolog	Kohlschutter–Tonz syndrome (KTZS)	226750
17p11.2	*PIGL*	N-acetylglucosaminyl-phosphatidylinositol de-N-acetylase	Coloboma, congenital heart disease, ichthyosiform dermatosis, mental retardation, and ear anomalies syndrome (CHIME)	280000
17q24.2–q24.3			Hypertrichosis terminalis, generalized, with our without gingival hyperplasia	135400
18q12.3	*SETBP1*	SET-binding protein	Schinzel–Giedion midface retraction synsdrome	269150
21q22.3	*RIPK4*	Receptor-interacting serine-threonine-protein kinase 4	Popliteal pterygium syndrome, lethal type	263650

[a]The *GJA1* gene was related only to Oculodentodigital dysplasia (ODDD) in Visinoni et al. [2009].

With the description of new syndromes and the better delineation of the conditions known, some syndromes initially described as different entities were considered variations of the same disorder. The opposite also occurs: descriptions considered as being the same clinical entity may be separated in two or more different conditions. This lumping and splitting of syndromes as well as the reports of previously undescribed conditions make the classifications dynamic and modifiable by incorporating the new knowledge. For instance, the TP63-related syndromes, ankyloblepharon-ectodermal defects-cleft lip and palate (AEC or Hay–Wells syndrome, OMIM 106260) and the Rapp–Hodgkin syndrome (OMIM 129400) are now considered as being different phenotypic expressions of the same condition [Prontera et al., 2008; Clements et al., 2010]. Furthermore, Prontera et al. [2011] suggested that ADULT

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EPIDERMOLYSIS BULLOSA: SIMPLEX, JUNCTIONAL AND DYSTROPHIC SUBTYPES AND KINDLER SYNDROME		
Subtype	**Inheritance**	**Defective protein(s)**
Suprabasal intraepidermal cleavage		
Acral peeling skin syndrome	AR	Transglutaminase 5
EB superficialis	Unknown	Unknown
Acantholytic EBS	AR	Desmoplakin; plakoglobin
Skin fragility syndromes		
Desmoplakin deficiency (skin fragility–woolly hair syndrome)	AR	Desmoplakin
Plakoglobin deficiency	AR	Plakoglobin
Plakophilin deficiency (skin fragility–ectodermal dysplasia syndrome)	AR	Plakophilin 1
Basal intraepidermal cleavage		
EBS, localized (formerly Weber–Cockayne)	AD	Keratins 5 and 14
EBS, generalized severe (formerly Dowling–Meara)		
EBS, generalized intermediate		
EBS with mottled pigmentation	AD	Keratin 5
EBS, migratory circinate		
EBS, AR-KRT14	AR	Keratin 14
EBS due to KLHL24	AD	Kelch-like protein 24
EBS with muscular dystrophy	AR	Plectin
EBS with pyloric atresia	AR	Plectin; $\alpha_6\beta_4$ integrin
EBS-Ogna	AD	Plectin
EBS, AR-BP230 deficiency	AR	Bullous pemphigoid antigen 1 (BP230)
EBS, AR-exophilin 5 deficiency	AR	Exophilin 5
Intra-lamina lucida cleavage		
JEB, generalized severe (formerly Herlitz)	AR	Laminin 332
JEB, generalized intermediate	AR*	Laminin 332; collagen XVII
JEB with pyloric atresia	AR	$\alpha_6\beta_4$ integrin
JEB, late onset	AR	Collagen XVII
JEB with respiratory and renal involvement	AR	α_3 integrin subunit
JEB, localized	AR	Collagen XVII; $\alpha_6\beta_4$ integrin
JEB, inversa	AR	Laminin 332
JEB-laryngo-onycho-cutaneous syndrome	AR	Laminin α3 chain (a isoform)
Sublamina densa cleavage		
DDEB, generalized	AD	Collagen VII
Other DDEB variants: acral, pretibial, pruriginosa, nails only		
DDEB, bullous dermolysis of the newborn		
RDEB, generalized severe (formerly Hallopeau–Siemens)	AR	
RDEB, generalized intermediate		
RDEB, inversa		
Other RDEB variants: localized, pretibial, pruriginosa, centripetalis		
RDEB, bullous dermolysis of the newborn		
Mixed cleavage planes		
Kindler syndrome	AR	Kindlin-1
*One case with AD inheritance has been reported.		

Table 32.1 Epidermolysis bullosa (EB): simplex, junctional and dystrophic subtypes and Kindler syndrome. Entities with a lighter background represent rare variants. AD, autosomal dominant; AR, autosomal recessive; EBS, epidermolysis bullosa simplex; DDEB, dominant dystrophic EB; RDEB, recessive dystrophic EB.

key component of the lamina lucida of the dermal–epidermal junction (see Ch. 28)[16]. In laryngo-onycho-cutaneous syndrome, the underlying mutations affect only the a isoform of the laminin α3 subunit. The milder generalized intermediate form of JEB (JEB-gen/intermed, previously known as JEB-non-Herlitz and generalized atrophic benign EB) results from mutations within the genes for either a subunit of laminin 332 or type XVII collagen. JEB with pyloric atresia, which is more common than EBS with pyloric atresia, is also caused by mutations within either of the two genes that encode the subunits of $\alpha_6\beta_4$ integrin. A newly recognized form of JEB associated with respiratory and renal involvement results from mutations in the integrin α_3 chain.

Dystrophic EB

Dystrophic EB (DEB) is transmitted in either an autosomal dominant or autosomal recessive manner and is caused by mutations in the type VII collagen gene. Dominant DEB (DDEB) results from dominant-negative mutations, typically a missense mutation that leads to substitution of

Table 6.2 Revised classification of inherited epidermolysis bullosa (EB)

Major EB type	Subtype	Gene defect	Target protein
Simplex (EBS)			
– suprabasal	Lethal acantholytic	*DSP*	Desmoplakin
	Plakophilin deficiency	*PKP1*	Plakophilin-1
	Superficialis	*?*	?
– basal	Localized (Weber–Cockayne)	*KRT5, 14*	K5, 14
	Dowling–Meara	*KRT5, 14*	K5, 14
	Other generalized (includes Koebner)	*KRT5, 14*	K5, 14
	EBS, with mottled pigmentation	*KRT5*	K5
	EBS, with muscular dystrophy	*PLEC1*	Plectin
	EBS, with pyloric atresia	*PLEC1, ITGA6, ITGB4*	Plectin, α6β4 integrin
	Autosomal recessive	*KRT14*	K14
	Ogna type	*PLEC1*	Plectin
	Migratory circinate	*KRT5*	K5
Junctional (JEB)			
– Herlitz	Herlitz	*LAMA3, LAMB3, LAMC2*	Laminin 5
– others	JEB, non Herlitz – generalized	*LAMA3, LAMB3, LAMC2,*	Laminin 5, type XVII collagen
	JEB, non Herlitz – localized	*COL17A1*	Type XVII collagen
	JEB, with pyloric atresia	*COL17A1*	α6β4 integrin
	JEB, inversa	*ITG176, ITGB4*	Laminin 5
	JEB, late onset	*LAMA3, LAMB3, LAMC2(?)*	?
	LOC syndrome	*?*	Laminin 332 (α3 chain)
		LAMA3	
Dystrophic (DEB)			
– dominant (DDEB)	DDEB, generalized	*COL7A1*	Type VII collagen
	DDEB, acral	*COL7A1*	Type VII collagen
	DDEB, pretibial	*COL7A1*	Type VII collagen
	DDEB, pruriginosa	*COL7A1*	Type VII collagen
	DDEB, nails only	*COL7A1*	Type VII collagen
	DDEB, bullous dermolysis of newborn	*COL7A1*	Type VII collagen
– recessive (RDEB)	RDEB, severe generalized	*COL7A1*	Type VII collagen
	RDEB, generalized other	*COL7A1*	Type VII collagen
	RDEB, inversa	*COL7A1*	Type VII collagen
	RDEB, pretibial	*COL7A1*	Type VII collagen
	RDEB, pruriginosa	*COL7A1*	Type VII collagen
	RDEB, centripetalis	*COL7A1*	Type VII collagen
	RDEB, bullous dermolysis of newborn	*COL7A1*	Type VII collagen
Kindler's syndrome	Kindler's syndrome	*KIND1*	Kindlin 1

ratories in any country will maintain stocks of these antibodies. The molecular defects, as currently known,[858] are listed in **Table 6.2**. A proposal to add a subgroup of hemidesmosomal cases, to include the plectin-deficient variant of epidermolysis bullosa simplex with muscular dystrophy, has not been incorporated into the most recent classification.[852,859,860] The intraepidermal/epidermolytic (simplex) group, which involves lysis of basal cells, will be considered first. Before doing so, brief mention will be made of the atypical nevi that may develop in all subsets of epidermolysis bullosa, particularly the recessively inherited variants.[861] The dermoscopic pattern may mimic melanoma but the absence of features such as irregular streaks, a blue-whitish veil, regression features, or black dots gives a distinctive dermoscopic pattern.[861]

Epidermolysis bullosa simplex

Epidermolysis bullosa simplex (EBS), one of the four major subgroups of epidermolysis bullosa (if Kindler's syndrome is accepted as the fourth category), is a mechanobullous disorder characterized by intraepidermal cleavage, usually through the basal layer of cells.[839,862] There are several distinct clinical variants.[863] Most cases have a mild clinical expression with blisters which heal without scarring, and an autosomal dominant inheritance.[864,865] Rare cases with an autosomal recessive inheritance and a more severe clinical course, sometimes with associated neuromuscular disease (particularly muscular dystrophy), have been reported.[866–868] Not all recessive forms have severe disease.[869,870] A prenatal diagnosis was made in the past using fetal skin biopsy samples.[822,871,872] Analysis of fetal DNA using chorionic villi or amniotic fluid cells is now the preferred technique.[873–876] Keratinocyte cell lines have been established for EBS, providing a potential source for in-vitro experiments using gene therapy.[877]

The molecular basis of most of the simplex subgroup resides in defects that disrupt the assembly, structure and/or function of the keratin intermediate filament skeleton of the basal keratinocyte – the coexpressed peptides keratin 5 and 14.[878–884] The keratin 5 (*KRT5*) gene maps to 12q13 and the keratin 14 (*KRT14*) gene to 17q12–q21. It appears that mutations in certain locations of the keratin genes (usually the ends of conserved α-helical rod domains) are more likely to produce the severe forms, such as the Dowling–Meara type (see below).[885] Twelve subtypes of EBS are recognized in the latest classification (see Table 6.2), which is based on the Report of the Third International Consensus Meeting held in 2007.[852] Only the four major subtypes of EBS will be considered in detail.

Other generalized type (includes Koebner variant)

The generalized (Koebner – OMIM 131900) clinical variant (now included in the 'other generalized type') is characterized by the devel-

Epidermolysis Bullosa

Table 23 Classification of epidermolysis bullosa

Major EB type	Major EB subtypes	Targeted protein(s)
EB simplex (EBS)	Suprabasal subtypes	
	• Lethal acantholytic EBS	Desmoplakin
	• Plakophilin-1 deficiency	Plakophilin-1
	• EBS superficialis (EBSS)	unknown
	Basal subtypes	
	• EBS, localized (Weber–Cockayne)	K5, K14
	• EBS, Dowling–Meara	K5, K14 – clumped tonofilaments on EM
	• EBS, other generalized (Koebner)	K5, K14
	• EBS with mottled pigmentation	K5
	• EBS with muscular dystrophy	Plectin
	• EBS with pyloric atresia (MC with JEB)	Plectin, α6β4 integrin
	• EBS, autosomal recessive	K14
	• EBS, Ogna variant	Plectin
	• EBS, migratory circinate	K5
Junctional EB(JEB)	JEB, Herlitz type	Laminin-332
	JEB, generalized non-Herlitz	Laminin-332; type XVII collagen
	JEB, localized non-Herlitz	Type XVII collagen
	JEB with pyloric atresia	α6β4 integrin
	JEB, inversa	Laminin-332
	JEB, late onset	unknown
	LOC (*l*aryngo-*o*nycho-*c*utaneous) syndrome (AKA -Shabbir's syndrome)	Laminin-332 (α3 chain)
Dominant dystrophic EB (DDEB)	DDEB, generalized	Type VII collagen
	DDEB, acral	
	DDEB, pretibial	
	DDEB, pruriginosa	
	DDEB, nails only	
	DDEB, bullous dermolysis of newborn (not always transient)	
Recessive dystrophic EB (RDEB)	RDEB, severe generalized (Hallopeau–Siemens)	Type VII collagen
	RDEB, generalized other (non-HS)	
	RDEB, inversa	
	RDEB, pretibial	
	RDEB, pruriginosa	
	RDEB, centripetalis	
	RDEB, bullous dermolysis of newborn (not always transient)	
Kindler syndrome		Kindlin-1

Table 8.1 List of the ichthyoses and related disorders

TYPES	SYNONYM
Non-syndromic ichthyoses	
Autosomal recessive congenital ichthyosis	ARCI
Lamellar ichthyosis	LI; ichthyosis congenita type 2
Congenital ichthyosiform erythroderma	CIE; nonbullous congenital ichthyosiform erythroderma; NBCIE, ichthyosis congenita type 1
Harlequin ichthyosis	HI; harlequin baby, harlequin fetus
Intermediate LI/CIE phenotypes	Non-classified ARCI
Self-healing collodion baby	SHCB
Bathing suit ichthyosis	BSI
Autosomal dominant congenital ichthyosis	
Autosomal dominant lamellar ichthyosis/ Autosomal dominant congenital ichthyosiform erythroderma	adLI; adCIE
Ichthyosis en confettis	Ichthyosis variegata, congenital reticular ichthyosiform erythroderma
Ichthyosis vulgaris	IV
Keratinopathic ichthyosis	KPI
Epidermolytic ichthyosis	EI; epidermolytic hyperkeratosis; EHK; bullous congenital ichthyosiform erythroderma of Brocq; bullous ichthyosis
Superficial epidermolytic ichthyosis	SEI; ichthyosis bullosa of Siemens; exfoliative ichthyosis
Mosaic epidermolytic ichthyosis	Mosaic EI, mosaic EHK; epidermal nevus of epidermolytic type
Annular epidermolytic ichthyosis	AEI
Ichthyosis Curth–Macklin	ICM; ichthyosis hystrix Curth–Macklin
KLICK syndrome	Keratosis Linearis with Ichthyosis Congenita and sclerosing Keratoderma
Loricrin keratoderma	LK; mutilating keratoderma with ichthyosis
Peeling skin syndrome	Keratolysis exfoliativa congenita; continual skin peeling; peeling skin syndrome type A/B, peeling skin disease, PSD
X-linked recessive ichthyosis	RXLI, steroid sulfatase deficiency
Syndromic ichthyoses	
ARC syndrome	Arthrogryposis, renal dysfunction, and cholestasis syndrome
CEDNIK syndrome	Cerebral dysgenesis, neuropathy, ichthyosis, and palmoplantar keratoderma
Chondrodysplasia punctata, X-linked dominant	Conradi–Hünermann–Happle syndrome
Ichthyosis hypotrichosis syndrome	HIS; congenital ichthyosis, follicular atrophoderma, hypotrichosis, and hypohidrosis
Ichthyosis–hypotrichosis–sclerosing cholangitis syndrome	IHSC syndrome; neonatal ichthyosis-sclerosing cholangitis syndrome; NISCH syndrome; ichthyosis, leukocyte vacuoles, alopecia, and sclerosing cholangitis
Ichthyosis prematurity syndrome	IPS
IFAP syndrome	Ichthyosis Follicularis, Atrichia, and Photophobia syndrome
Infantile Gaucher disease	Gaucher disease type II; GD II; Gaucher disease of the acute neuronopathic type
KID syndrome (including HID syndrome)	Keratitis–Ichthyosis-like–Deafness syndrome Ichthyosiform erythroderma, corneal involvement, and deafness, autosomal dominant (hystrix-like ichthyosis-deafness syndrome; ichthyosis hystrix gravior, type Rheydt)
MEDNIK syndrome	Mental retardation, Enteropathy, Deafness, peripheral Neuropathy, Ichthyosis, and Keratodermia syndrome
Multiple sulfatase deficiency	MSD
Netherton syndrome/ichthyosis linearis circumflexa	NS; NTS; Comèl–Netherton syndrome
Neutral lipid storage disease with ichthyosis	Chanarin–Dorfman syndrome; triglyceride storage disease with impaired long-chain fatty acid oxidation; ichthyosiform erythroderma with leukocyte vacuolization
Refsum disease	Adult Refsum disease; heredopathia atactica polyneuritiformis; phytanic acid oxidase deficiency; hereditary motor and sensory neuropathy IV; HMSN4
Rhizomelic chondrodysplasia punctata (RCDP1)	Rhizomelic chondrodyslplasia type 1; chondrodystrophia calcificans punctata
Sjögren–Larsson syndrome	SLS
Trichothiodystrophy	Tay syndrome; (P)IBIDS syndrome: (Photosensitivity)–Ichthyosis-TTD; Brittle hair–Impaired intelligence–Decreased fertility–Short stature syndrome
Related disorders	
CHILD syndrome	Unilateral CIE
CHIME syndrome	Ocular Colobomas, congenital Heart disease, early-onset Ichthyosiform dermatosis, Mental retardation and Ear anomalies (conductive hearing loss)/Epilepsy; Zunich–Kaye syndrome; Zunich neuroectodermal syndrome
Desmons syndrome	Ichthyosiform erythroderma, corneal involvement, and deafness; autosomal recessive
Erythrokeratodermia variabilis	EKV, erythrokeratodermia figurate variabilis Mendes da Costa
Ichthyosis, hepatosplenomegaly, and cerebellar degeneration	
Ichthyosis–mental retardation syndrome with large keratohyalin granules	
Keratosis spinulosa delcalvans	KFSD
Neu-Laxora syndrome	NLS
Acquired ichthyosis	

Table 1 Nonsyndromic ichthyoses

Disease	Mode of inheritance	Gene(s)
Common ichthyoses		
- Ichthyosis vulgaris [146700]	Autosomal semidominant	*FLG*
- X-linked ichthyosis – nonsyndromic [308100]	X-linked	*STS*
Autosomal-recessive congenital ichthyoses		
- Harlequin ichthyosis		
o ARCI4B [242500]	AR	*ABCA12*
- Lamellar ichthyosis and/or **- Congenital ichthyosiform erythrodermia and/or** **- Self-improving congenital ichthyosis**		
o ARCI1 [242300] o ARCI2 [242100] o ARCI3 [606545] o ARCI4A [601277] o ARCI5 [604777] o ARCI6 [612281] o ARCI8 [613943] (late onset) o ARCI9 [615023] o ARCI10 [615024] o ARCI11 [613943] (see also Table 2) o ARCI12 [605848] o ARCI13 [609769] o ARCI14 [604125]	AR	*TGM1* *ALOX12B* *ALOXE3* *ABCA12* *CYP4F22* *NIPAL4* *LIPN* *CERS3* *PNPLA1* *ST14* *CASP14* *SDR9C7* *SULT2B1*
- Self-healing collodion baby		
o ARCI1 [242300]	AR	*TGM1*
- Bathing suit ichthyosis		
o ARCI1 [242300]	AR	*TGM1*
	AD	*ASPVR1*
Keratinopathic ichthyosis		
o Epidermolytic ichthyosis [113800]	AD	*KRT1/KRT10*
o Superficial epidermolytic ichthyosis [146800]	AD	*KRT2*
- KPI variants		
o Annular epidermolytic ichthyosis [607602]	AD	*KRT1/KRT10*
o Ichthyosis Curth–Macklin [146590]	AD	*KRT1*
o Autosomal-recessive epidermolytic ichthyosis [113800]	AR	*KRT10*
o Congenital reticular ichthyosiform erythrodermia/ichthyosis with confetti [609165]	AD	*KRT10/KRT1*
o Epidermolytic nevi [113800][a]	Somatic mutations	*KRT1/KRT10*
Peeling skin syndromes – generalized		
- Peeling skin disease type B [270300]	AR	*CDSN*
- Peeling skin disease type A [610190] [616284]		*CHST8/FLG2*
- Exfoliative ichthyosis [607936] - Exfoliative ichthyosis [617115]		*CSTA* *SERPINB8*
Other nonsyndromic ichthyoses		
- Autosomal dominant lamellar ichthyosis [146750]	AD	ASPVR1
- Keratosis linearis–ichthyosis congenita keratoderma [601952]	AR	*POMP*
- Loricrin keratoderma [604117]	AD	*LOR*
- Erythrokeratodermia variabilis [133200]	AD	*GJB3/GJB4*

AD autosomal dominant, *AR* autosomal recessive
[a]May indicate a gonadal mosaicism, which can cause generalized epidermolytic ichthyosis in the offspring generation. OMIM in [ ], http://www.ncbi.nlm.nih.gov/omim

their environment. The squamous epithelium consists of keratinocytes, whose degree of differentiation is distinguished functionally and morphologically by several histological layers (Figs. 1 and 2).

3.1 Stratum Basale (Basal Cell Layer)

Here the cells are aligned vertically. Within the layer and immediately above it (stratum suprabasale) mitosis takes

Table 2 Syndromic ichthyoses

Disease	Mode of inheritance	Gene(s)
X-linked ichthyosis syndromes		
- X-linked ichthyosis, syndromic [308100, 300500, 300533]	X-linked	*STS* (and others)
- Ichthyosis follicularis alopecia photophobia syndrome [308205]		*MBTPS2*
- Conradi–Hünermann–Happle syndrome (CDPX2) [302960]		*EBP*
Autosomal ichthyosis syndromes with prominent hair abnormalities		
- Netherton syndrome [256500]	AR	*SPINK5*
- Ichthyosis hypotrichosis syndrome [602400]		*ST14*
- Neonatal ichthyosis-sclerosing cholangitis/ichthyosis hypotrichosis sclerosing cholangitis syndrome [607626]		*CLDN1*
Autosomal ichthyosis syndromes with prominent neurological signs		
- Keratitis ichthyosis deafness syndrome [148210]	AD	*GJB2* (*GJB6*)
- Refsum syndrome (HMSN4) [266500]	AR	*PHYH/PEX7*
- Multiple sulfatase deficiency [272200]		*SUMF1*
- Gaucher syndrome type 2 [230900]		*GBA*
- Sjögren–Larsson syndrome [270200]		*ALDH3A2*
- Trichothiodystrophy/Tay syndrome o TTD1 [601675]	AR	*ERCC2/XPD*
o TTD2 [616390]		*ERCC3/XPB*
o TTD3 [616395]		*GTF2H5/TFIIH*
- Cerebral dysgenesis–neuropathy–ichthyosis–palmoplantar keratoderma syndrome [609528]	AR	*SNAP29*
- Mental retardation–enteropathy–deafness–neuropathy–ichthyosis–keratoderma syndrome [609313]		*AP1S1*
- Arthrogryposis–renal dysfunction–cholestasis syndrome [208085]		*VPS33B*
- Ichthyosis–spastic quadriplegia–mental retardation syndrome [614457]		*ELOVL4*
- Coloboma–congenital heart disease–ichthyosiform dermatosis–mental retardation–ear anomalies syndrome [280000]		*PIGL*
Autosomal ichthyosis syndromes with other prominent findings		
- Desmons syndrome [600157]		*AP1B1*
- Ichthyosis prematurity syndrome [608649]	AR	*SLC27A4* (*FATP4*)
- Severe dermatitis–multiple allergies–metabolic wasting syndrome [615508]		*DSG1* (see also *DSP*)
- Chanarin–Dorfman syndrome/neutral lipid storage disease with ichthyosis [275630]		*ABHD5*
- Neu–Laxova syndrome 1 [256520]		*PHGDH*
- Neu–Laxova syndrome 2 [616038]		*PSAT1*

AD autosomal dominant, *AR* autosomal recessive, *CDPX2* chondrodysplasia punctata type 2, *HMSN4* hereditary motor and sensory neuropathy type 4; OMIM in [ ], http://www.ncbi.nlm.nih.gov/omim

place regularly. The migration and differentiation of the cells from the stratum basale to the stratum granulosum takes place within about 14 days. It takes another 14 days for the corneocytes to exfoliate, resulting in a total epidermal turnover of approximately 28 days, which can be considerably shortened depending on the disease or skin condition.

The intermediate filaments of the epidermis, i.e., cytoskeletal structures of 10 nm, consist mainly of keratins. At least 54 functionally relevant keratin genes can be differentiated in terms of tissue and in the skin and its appendages (Table 3). Type I keratins differ from type II keratins in size (40–57.5 kDa and 53–67 kDa respectively) and charge (acidic or alkaline–neutral). Equimolar amounts of keratin types I and II polymerize with each other by spontaneous parallel arrangement to keratin dimers, which in turn polymerize by antiparallel arrangement to protofilaments. At the end electron density keratin filaments are formed from protofibrils with a diameter of 5–8 nm, which together form sound fibrils with a diameter of about 25 nm. The basal keratinocytes mainly produce keratin 14 and 5, as well as type IV collagen, laminins, and integrins. The basement membrane is thus a product of the epidermis.

In addition to keratins, lipids are of great importance for the structure of the epidermis. Besides the neutral lipids (glycerol phosphatides, triglycerides, free fatty acids), complex sphingolipids (ceramides and glycosylceramides) occur in the epidermis, which are otherwise only found in the nervous system. In the basal epidermis the phospholipids form the largest lipid fraction in the cell membrane. During the keratinization process, a considerable increase in neutral

may require referral for cardiac, audiologic, ophthalmologic, or dental evaluation.

A skin biopsy for histologic examination may provide additional diagnostic clues (Table 58.4). The molecular defect underlying PPKs may involve epidermal proteases as well as proteins of the keratinocyte cytoskeleton, cornified envelope, desmosome (see Fig. 56.8), or gap junction (Fig. 58.4, Table 58.5); the known underlying genetic mutations are summarized in Table 58.6[5,6]. Genetic analysis is sometimes helpful to establish the specific diagnosis, facilitate identification of affected family members, and enable prenatal diagnosis.

PRIMARY PALMOPLANTAR KERATODERMAS (PPKS)	
Disorder	**Additional features**
Hereditary PPKs	
Diffuse PPK, isolated	
Vörner–Unna–Thost	Epidermolytic
Nagashima, Kimonis	Non-epidermolytic
Bothnia	White/spongy upon water exposure; non-epidermolytic
"Greither"	Transgrediens & progrediens; variable pathology
Mal de Meleda (including Gamborg Nielsen)	Mutilating, frequent superinfections; non-epidermolytic
Diffuse PPK with associated features	
Hearing impairment:	
Vohwinkel syndrome	Mutilating, "honeycomb" PPK; starfish keratoses on knuckles
Bart–Pumphrey syndrome	Knuckle pads, leukonychia
PPK due to connexin 26 defect	"Honeycomb" PPK
Mitochondrial PPK	
High risk of acral SCC:*	
Huriez syndrome	Scleroatrophy
PPK with sex reversal	Female-to-male sex reversal
Hypotrichosis & nail dystrophy:	
Clouston syndrome	Pebbled/grid-like acral papules
Odonto-onycho-dermal dysplasia	Hypodontia, facial telangiectasias/erythema
Schöpf–Schulz–Passarge syndrome	Hidrocystomas, other adnexal neoplasms, hypodontia
Keratoderma–hypotrichosis–leukonychia totalis	Total leukonychia
Mutilating:*	
Olmsted syndrome	Periorificial/intertriginous keratotic plaques
Papillon–Lefèvre syndrome**	Periodontitis, pyogenic infections, psoriasiform plaques
Loricrin keratoderma	Mild generalized ichthyosis, "honeycomb" PPK
KLICK	Keratosis linearis, ichthyosis congenita, sclerosing keratoderma
Vohwinkel syndrome (see above)	
Mal de Meleda (see above)	
Cardiomyopathy:	
Naxos disease***	Woolly hair, arrhythmias in adolescence
Focal PPK, isolated	
Brünauer–Fuhs–Siemens, Wachters	Striate on palms, nummular on soles; see Table 58.6 for genetic subtypes
Focal PPK with associated features	
Pachyonychia congenita	Painful soles, dystrophic nails, oral leukokeratosis, steatocystomas/other cysts
Focal palmoplantar and gingival keratosis	Painful soles, gingival leukokeratosis; epidermolytic
Howel–Evans syndrome	Esophageal carcinoma
Richner–Hanhart syndrome (oculocutaneous tyrosinemia)	Dendritic keratitis, corneal ulcers, intellectual disability, painful keratosis
Carvajal syndrome***	Striate on palms; woolly hair, dilated cardiomyopathy; occasionally skin fragility, nail dystrophy, and hypodontia
PPK and woolly hair	Striate on palms; woolly hair, hypotrichosis, leukonychia

*Acral SCC can also develop in patients with mutilating PPK.
**_Haim–Munk syndrome_ is an allelic disorder that also features arachnodactyly, atrophic nail changes, and acro-osteolysis.
***The PPK in Naxos disease is occasionally focal/striate, overlapping with Carvajal syndrome.

Table 58.1 Primary palmoplantar keratodermas (PPKs). More common disorders are in **bold** font. Other genodermatoses that feature PPK are listed in Table 58.2. Of note, the pattern can be different on the hands and feet, e.g. diffuse on the feet and focal on the hands. Progrediens means progressive, referring to the tendency to involve additional sites (e.g. the elbows and knees) over time. Mutilating refers to the development of digital constrictions and possible autoamputation (pseudoainhum; see Table 58.3). SCC, squamous cell carcinoma.

Continued

PRIMARY PALMOPLANTAR KERATODERMAS (PPKS)	
Disorder	**Additional features**
Punctate PPK, isolated	
Buschke–Fischer–Brauer (type 1 punctate PPK) Spiny keratoderma (type 2 punctate PPK) Marginal papular keratoderma: acrokeratoelastoidosis, focal acral hyperkeratosis (type 3 punctate PPK) **Punctate keratoses of the palmar creases** Porokeratosis punctata palmaris et plantaris	
Punctate PPK with associated features	
Cole disease	Guttate hypopigmentation, calcinosis cutis
PLACK syndrome	*P*eeling skin, *l*eukonychia, *c*heilitis, *k*nuckle pads
Acquired PPKs	
Keratoderma climactericum **Aquagenic PPK** Pityriasis rubra pilaris Associated with hypothyroidism/myxedema Associated with cancer Associated with lymphedema Arsenical keratoses Drug-induced keratoderma (e.g. lithium, verapamil, venlafaxine, glucan, tegafur, capecitabine, imatinib, sorafenib, sunitinib) Induced by dioxin or halogenated herbicides Associated with dermatomyositis: pityriasis rubra pilaris-like (Wong) type, "mechanic's hands" in antisynthetase syndrome	

Table 58.1 Primary palmoplantar keratodermas (PPKs). (cont'd)

Fig. 58.2 Focal palmoplantar keratoderma. A Striate type with linear hyperkeratosis on the palm overlaying flexor tendons. **B** Areata type on the soles. *B, Courtesy, Alfons Krol, MD, and Dawn Siegel, MD.*

HEREDITARY KERATODERMAS

DIFFUSE PALMOPLANTAR KERATODERMA WITHOUT ASSOCIATED FEATURES (ISOLATED, NON-SYNDROMIC)

Diffuse Epidermolytic Palmoplantar Keratoderma (EPPK)

Synonyms: ■ Unna–Thost disease ■ Vörner disease ■ Vörner–Unna–Thost type PPK ■ Keratosis palmoplantaris diffusa Vörner–Unna–Thost

History

In 1880, Thost described a family with diffuse non-transgrediens PPK. This was followed by Unna's description of a clinically identical, autosomal dominant PPK in two families. Although "Unna–Thost type" has been used to designate non-epidermolytic PPK (NEPPK), in 1992, Küster et al.[7] reviewed the family described by Thost and found the histologic features of epidermolytic hyperkeratosis, which characterize the "Vörner type" of PPK. Subsequently, after keratin mutations were reported in epidermolytic PPK (EPPK), identical mutations were found in the descendants of the family studied by Thost[8]. This confusion underscores the need for careful histologic examination and sometimes multiple biopsy specimens to identify epidermolytic hyperkeratosis.

Epidemiology

EPPK is likely the most common form of diffuse keratoderma, with an estimated incidence of ≥4.4 per 100 000 in Northern Ireland[9].

Pathogenesis

Mutations in *KRT1* and *KRT9* underlie diffuse EPPK. The expression of *keratin 9* is limited to the suprabasal cell layers of palmar and plantar skin. In patients with diffuse EPPK, the majority of keratin 9

HERITABLE FORMS OF CUTIS LAXA AND RELATED CONDITIONS					
Disorder	**Inheritance**	**Gene**	**Protein**	**Distribution of cutis laxa**	**Additional features**
Heritable forms of cutis laxa					
ADCL	AD[†]	*ELN* *FBLN5*	Elastin Fibulin 5	Generalized	• Cardiac valve abnormalities, aortic dilatation (variable) • Emphysema (uncommon) • Hernias[‡]
ARCL type 1B ARCL type 1A	AR	*FBLN4 (EFEMP2)* *FBLN5*	Fibulin 4 Fibulin 5	Generalized	• Arterial or aortic tortuosity/aneurysms (1B) and stenosis • Emphysema (more severe in 1A) • Bladder diverticula, hernias[‡] • In 1B: joint laxity, arachnodactyly, fractures, micrognathia
ARCL type 1C (Urban–Rifkin–Davis syndrome)	AR	*LTBP4*	Latent TGF-β binding protein-4	Generalized	• Pulmonary malformations/ emphysema (severe, often fatal) • Pulmonary arterial stenosis, valvular defects • GI and GU malformations (diverticula, stenoses) • Micrognathia, large fontanelles • Joint laxity, hypotonia
ARCL type 2A (Debré type; allelic to wrinkly skin syndrome)	AR	*ATP6V0A2*	Vacuolar H^+-ATPase V0, subunit A2	Generalized; improves over time	• Translucent skin with prominent veins (especially 2B, DBS) • Ocular abnormalities: strabismus & high myopia (2A); corneal opacities & cataracts (DBS, PADCL) • Facial dysmorphism[§]; triangular face with more progeroid appearance (2B, DBS); large fontanelles • Microcephaly, partial pachygyria (2A), absent corpus callosum (2B) • Intellectual disability; neurologic degeneration (2A); dystonia/athetosis (DBS) • Congenital hip dysplasia, joint laxity, hypotonia, scoliosis, short stature; osteopenia (2B, DBS) • Cranial arterial tortuosity (DBS-A, PADCL) • O- and N-glycosylation defect (2A[‖])
ARCL type 2B De Barsy syndrome (DBS) B (ARCL type 3B)	AR	*PYCR1*	Pyrroline-5-carboxylate reductase 1	Generalized; may favor dorsal hands/feet (2B) or improve over time; occasionally fat pads (e.g. on buttocks; DBS-A)	
Progeroid ADCL (PADCL) De Barsy syndrome (DBS) A (ARCL type 3A)	AD AR	*ALDH18A1*	Pyrroline-5-carboxylate synthetase		
ARCL types 2C and 2D	AR	*ATP6V1E1* (2C) *ATP6V1A* (2D)	Vacuolar H^+-ATPase V1, subunits E1 and A	Generalized with sparse or subcutaneous fat; large skin folds on extremities, fat pads on buttocks; may improve over time (2D)	• Pneumothoraces • Cardiac abnormalities, aortic dilation • "Mask-like" triangular face, hypotonia; variable hip dysplasia, marfanoid habitus • Neurodevelopmental abnormalities (2D)
Gerodermia osteodysplastica (geroderma osteodysplasticum)	AR	*GORAB*	Golgin, RAB6-interacting	Primarily on dorsal hands/feet	• Malar hypoplasia, prognathism, oblique furrow lateral to eyebrows • Joint laxity • Osteoporosis, fractures, dwarfism
Macrocephaly, alopecia, CL, and scoliosis (MACS)	AR	*RIN2*	RAS and RAB interactor 2	Generalized, most pronounced on face	• Sparse hair • Gingival hypertrophy, facial dysmorphism • Macrocephaly • Scoliosis, joint laxity

Table 97.6 Heritable forms of cutis laxa (CL) and related conditions. In addition to ARCL types 2A and 2C, CL can occur in other congenital disorders of glycosylation, e.g. due to mutations in a component of oligomeric Golgi complex 7 (*COG7*; multiple-system malformations) or steroid 5-α-reductase 3 (*SRD5A3*; also ichthyosis, ocular colobomas, and midline brain malformations). Transaldolase deficiency can present with neonatal CL (generalized or of the forehead) that improves over time as well as hypertrichosis, hepatosplenomegaly, hemolytic anemia, and GU malformations. "Neonatal CL with a marfanoid phenotype" was described in an infant who lacked immunohistochemical evidence of laminin-β1 in the skin. AD, autosomal dominant; *ALDH18A1*, aldehyde dehydrogenase 18 family, member A1; AR, autosomal recessive; *EFEMP2*, EGF-containing fibulin-like extracellular matrix protein 2; GI, gastrointestinal; GU, genitourinary; TGF, transforming growth factor; X-R, X-linked recessive.

HERITABLE FORMS OF CUTIS LAXA AND RELATED CONDITIONS					
Disorder	**Inheritance**	**Gene**	**Protein**	**Distribution of cutis laxa**	**Additional features**
Heritable forms of cutis laxa					
Occipital horn syndrome	X-R	*ATP7A*	ATPase, Cu^{2+}-transporting, α-polypeptide	Generalized, most pronounced on face and neck	• Easy bruising, coarse hair (variable) • Arterial tortuosity • Chronic diarrhea; bladder diverticula • Long face, high forehead, hooked nose • Developmental delay • Wedge-shaped occipital calcifications • Joint laxity
Related heritable conditions that feature lax and/or redundant skin					
Arterial tortuosity syndrome	AR	*SLC2A10 (GLUT10)*	Solute carrier family 2, member 10 (a glucose transporter)	Generalized; soft, hyperextensible skin has also been described	• Telangiectasias on cheeks (variable) • Arterial tortuosity • Long face, micrognathia, high-arched palate • Joint laxity and contractures
Larsen-like syndrome	AR	*B3GAT3*	Glucuronosyltransferase I	Wrinkled acral skin	• Joint dislocations, craniofacial dysmorphism, short stature • Congenital heart defects
Costello syndrome	AR	*HRAS*	HRAS	Variable, especially on hands and feet	• Deep palmoplantar creases, acanthosis nigricans, periorificial papillomas • Curly or sparse hair, coarse facies • Cardiac malformations • Mental and growth retardation • Risk of malignancies, e.g. rhabdomyosarcoma
Donohue syndrome ("leprechaunism"; see Ch. 101)	AR	*INSR*	Insulin receptor	Variable	• Generalized lipodystrophy, hypertrichosis, acanthosis nigricans • Insulin resistance • Distinctive "elfin" facies • Severe growth retardation
Finnish-type amyloidosis	AD	*GSN*	Gelsolin	Generalized	• Corneal dystrophy • Polyneuropathy
Cranioectodermal dysplasia (Sensenbrenner syndrome)	AR	*IFT122* *IFT43*	Intraflagellar transport proteins 122 and 43	Generalized; redundant skin folds at neck, ankles, and wrists	• Sparse, fine hair; short, broad nails • Narrow thorax, short proximal limbs, joint laxity, brachydactyly, dolichocephaly, dysmorphic facies, dental anomalies • Nephronophthisis, hepatic fibrosis, retinal dystrophy
		WDR35 *WDR19*	WD repeat domain 35 and 19		
Circumferential skin creases, Kunze type	AD, AR	*MAPRE2*	Microtubule-associated protein RP/EB family member 2	Multiple circumferential creases/folds of skin on the extremities (see Fig. 97.12) > neck > trunk (excess rather than lax skin); may resolve over time	• Cleft palate, short stature, dysmorphic facies (e.g. epicanthal folds, small eyes), overfolded helices • Intellectual disability
	AD	*TUBB*	Tubulin-β class I		
Nonsyndromic "Michelin tire baby"	AD	?	?		• Affected skin may have excess fat or smooth muscle, and the latter is often associated with hypertrichosis; elastic fibers are usually normal (fragmentation noted in only one report)

†An AR form of cutis laxa due to a homozygous elastin mutation has been reported in two related families.
‡E.g. inguinal, umbilical, diaphragmatic.
§Features may include a broad forehead and midfacial hypoplasia as well as findings related to the lax skin (e.g. down-slanting palpebral fissures, long philtrum).
‖Can be assessed via analysis of the glycosylation of serum transferrin ± apolipoprotein C-III.

Table 97.6 Heritable forms of cutis laxa (CL) and related conditions. (cont'd)

particularly pulmonary complications, can cause considerable morbidity and mortality[64]. Association with congenital hemolytic anemia has also been reported[65]. The cutaneous and extracutaneous features of specific types of heritable CL are summarized in Table 97.6.

Generalized acquired CL without an identifiable trigger may be clinically indistinguishable from milder heritable forms of CL, e.g. late-onset autosomal dominant CL. Generalized or localized forms of acquired CL can also occur in association with a plasma cell dyscrasia (see Fig. 97.10B), as a consequence of drug intake (particularly penicillamine), and subsequent to a systemic or cutaneous inflammatory disorder (Table 97.7). In the latter situation, the inflammatory reaction is followed by progressive sagging of the skin, which can become exten-

- Mitral and aortic valve prolapse/dilatation and regurgitation
- Cystic medionecrosis of the aorta
- Neurologic features include macrocephaly, hydrocephalus, syringomyelia, and basilar invagination

- Variable prognosis depending on disease type and severity:
  - Types I and IV: normal life span
  - Type II: **death in perinatal period**
  - Type III: increased mortality in 3rd/4th decade due to **respiratory failure** (secondary to kyphoscoliosis) or head trauma

Ehlers-Danlos syndrome (EDS)

- Heterogeneous group characterized by abnormal collagen structure and/or function within the skin, joints, and vasculature (Table 4-13)
- Classical EDS (most common subtype)
  - May deliver/be delivered pre-term as a result of early rupture of fetal membranes; normal life span
  - Mucocutaneous manifestations
    - Velvety, soft, and doughy consistency of skin
    - **Marked hyperextensibility of skin**
    - **Poor wound healing ("cigarette paper" scars)**

Table 4-13. Ehlers-Danlos Syndrome Classification

Type	Genes	Inheritance	Skin Findings	Joint Changes	Other
Classical (formerly **Type I**, "gravis" and **Type II**, "mitis")	***COL5A1***, ***COL5A2***	AD	Hyperextensibility Easy bruising Fragile skin Widened atrophic ("**fish mouth**" or "**cigarette paper**") scars **Molluscoid pseudotumors** (overlying extensor joints and pressure points) Spheroids	Hypermobility and joint dislocations	**(+)Gorlin's sign** Absence of lingual frenulum
Hypermobility (formerly **Type III**)	*TNXB*	AD/AR	Mild	**Hypermobility** Chronic joint pain + arthritis Recurrent **dislocations and subluxations**	Deficiency of **tenascin X**
Vascular (formerly **Type IV**)	***COL3A1***	AD	Thin, translucent skin Extensive bruising Early varicosities (can **visualize veins under skin** easily) Acrogeria	Small joint hypermobility	Mnemonic: "IV = blood vessels" **Rupture of bowel, uterus, or arteries** Most **life-threatening** form
Kyphoscoliotic (formerly **Type VI**)	Lysyl hydroxylase 1 (*PLOD1*)	AR	Mild	Hypermobility **Severe scoliosis**	Severe muscle hypotonia **Ruptured globe, blindness, retinal detachment, or keratoconus** Marfanoid Osteopenia Ascorbic acid supplementation may help
Arthrochalasic (formerly **Types VIIa and VIIb**)	*COL1A1*, *COL1A2*	AD	Mild	**Most severe hypermobility** with recurrent subluxations/ dislocations (much more severe than Hypermobility type)	**Congenital hip dislocation** Short stature
Dermatosparaxis (formerly **Type VIIc**)	Procollagen N-proteinase (***ADAMTS2***)	AR	Severe fragility **Sagging**, **redundant skin**, and bruising	Mild	Umbilical/inguinal hernias Premature rupture of fetal membranes
Periodontitis type (formerly **Type VIII**)	Unclear	AD	Hyperextensible skin with scarring (esp. pretibial) and bruising	Mild	**Severe periodontitis → teeth loss**

(Adapted from Paller S, Mancini AJ. Hurwitz Clinical Pediatric Dermatology, 4th Ed. Elsevier. 2011)

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Dermatology diseases Index

FIRST INDEX OF SKIN DISEASES IN THE WORLD

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